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HYPOSPADIAS REPAIR

**with the transverse inner
preputial island flap technique**

J.D.M. de Vries

**HYPOSPADIAS REPAIR
WITH THE TRANSVERSE INNER
PREPUTIAL ISLAND FLAP TECHNIQUE**

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PROEFSCHRIFT

ter verkrijging van de graad van doctor in de geneeskunde
aan de Katholieke Universiteit te Nijmegen
op gezag van de rector magnificus Prof. dr. J.H.G.I. Giesbers,
volgens het besluit van het college van decanen in het openbaar te verdedigen
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door

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geboren te Rotterdam

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This study of which the results are presented in this thesis was performed at the Department of Urology of the Rijksuniversiteit and of the Maria Middelaers Hospital, both in Ghent Belgium (Chief and chairman Prof dr W A de Sy) and at the Department of Urology of the Catholic University, Nijmegen, the Netherlands (Chief and chairman Prof dr F M J Debruyne). The operations were performed by Prof dr W A de Sy in Belgium and by the author in the Netherlands.

*Nulla autem est alia pro certo noscendi via,
nisi quam plurimas et morborum et dissectionem
historias, tum aliorum, tum proprias collectas
habere et inter se comparare*

Giov Batt MORGAGNI (1682-1771)

*„De sedibus et causis morborum
per anatomen indagatis libri quinque“*

To my Parents
Maria
Gerdjan
Franck
Laurens

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Hypospadias is a relatively common malformation of the male genital system, with an estimated incidence of one percent in certain areas (Opitz 1985). According to recent investigations the incidence seems to increase (Kallen and Winberg 1982, Czeizel 1985, Matlai and Beral 1985). The only possible cure for this malformation is a surgical correction. Until now more than two hundred corrective procedures have been described. A critical reappraisal of these methods reveals, as expected, that only a few are actually new. We completely agree with Marberger and Pauer (1981), who stated: "It seems unlikely, that new operative techniques for hypospadias repair can be invented since over two hundred methods have been applied so far". However, in the last decades considerable progress has been made in the understanding of the ontogenesis of this malformation. This insight has refined the operative procedures and the introduction of new suture and wound-dressing material has further improved the outcome.

Confronted with the results of the transverse inner preputial island flap technique as performed by Duckett in Philadelphia and by Ransley in London, we changed from the multi stage correction to this type of one stage repair. We adopted this technique since it made a correction possible in one session for the majority of hypospadiac patients. Furthermore a correction can be performed at an early age, which has the advantage that several psychological sequelae of any operative correction of the external genitalia can be prevented. We were able to show by experimental studies, that the vasculature of the prepuce allows the preparation of a well vascularized pedicled island flap.

In this study we retrospectively analysed the results of this method of hypospadias repair and compared our results with data from the literature.

The hypospadiac entity

1.1 Morphology

The hypospadiac entity is characterized by several typical anatomical features:

- 1 The ectopy of the urethral orifice
- 2 The lack of a frenulum and the asymmetrical development of both the raphe and the prepuce
- 3 The chordee, causing a bending of the penis

1.1.1 The ectopy of the urethral orifice

The urethral orifice can be located at the perineum, the scrotum, the peno-scrotal corner, the ventral penile body or at the coronal-glandular area, depending on the moment (early to late) at which the urethral plate stops to form a tube (fig. 1). This event takes place in the early fetal period (see page 15).

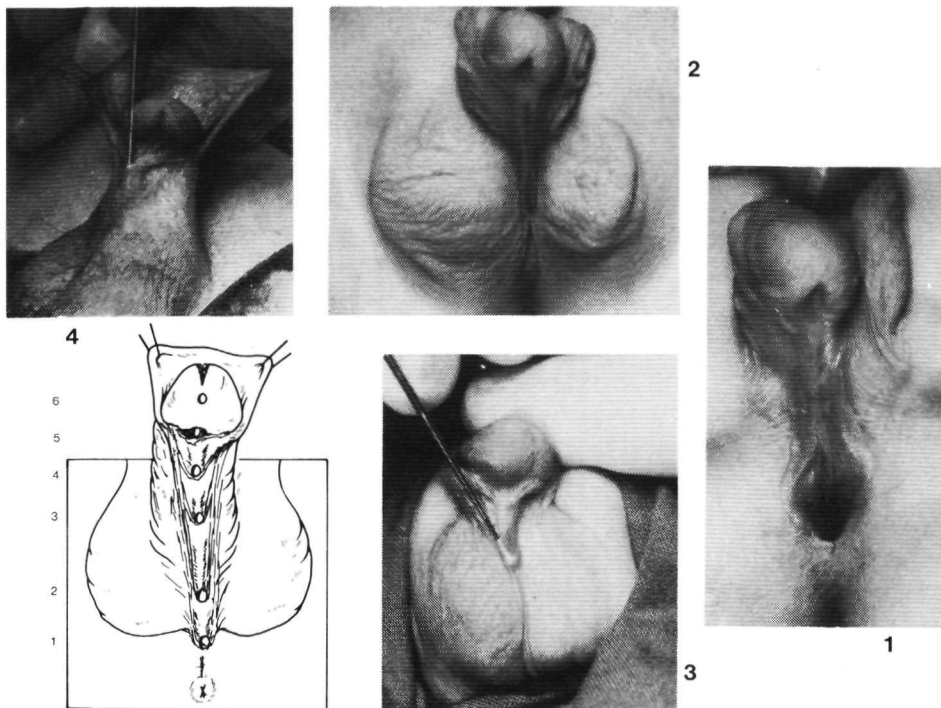


Fig. 1: The possible position of the meatus in hypospadias
After Marberger and Altwein (1983)

The ectopic meatus is very often stenotic. This stenosis can be so severe that it causes micturation problems and a meatotomy has to be performed as an emergency procedure in early childhood. This problem is mostly seen in more distally located orifices.

1.1.2 The lack of a frenulum and the asymmetrical development of both the raphe and the prepuce

A frenulum is always lacking. This is due to the fact, that epithelial lining at both sides of the urethral plate at the level of the ectopic orifice, do not fuse. Therefore distally of this point there is no formation of a raphe. Surprisingly little attention has been given to the shape and type of splitting of the raphe. Only Ombredanne (1932), Glenister (1954) and van der Meulen (1964) discussed this phenomenon.

The raphe can split up symmetrically in two parts, creating a "V" shaped skin defect. It can also be located completely at one side. This gives sometimes a torsion of the skin with the impression of a torsion of the penis itself (fig. 2).



Fig. 2: Asymmetrical epithelial lining causing torsion of the penile skin

In general the raphe ends, after splitting, at the dorso- lateral aspect of the penis in small epithelial cones: the so called dogears (fig. 3).

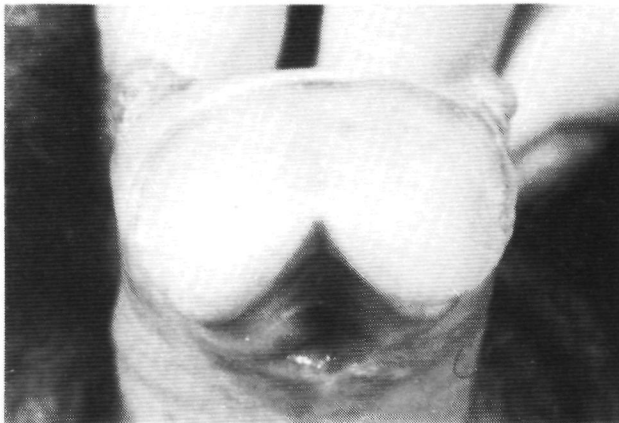


Fig. 3: Asymmetrical epithelial lining, forming a V-shaped defect and "dogears"

1.1.3 The chordee (bending of the penis)

In the majority of the hypospadiac cases a bending of the penis is present. This bending can be mild, moderate or severe.

The curvature is always concave at the ventral side. The glans is mostly mal-positioned as well. The coronal sulcus is not oriented in a normal ventro-dorsal position of 180 degrees to the long axis of the corpora but makes an angle between 15 and 45 degrees with this axis: the glandular tilt.

An other change in the anatomical appearance that can be observed, is the dystopy of the penis.

This relatively rare phenomenon (peno-scrotal transposition, engulfment or webbed penis) is mostly observed in more severe hypospadias. It looks, as if the scrotal halves fuse cranially of the penile base.



Fig. 4: Transposition in a severe hypospadiac case

It can however also be observed in sexually normally developed male children. So a direct relationship with the hypospadiac entity does not exist. It sometimes can cause embarrassment in the sexual assignment of the newborn, especially if it is accompanied by a scrotal bipartition.

1.2 Classification

The mostly used classification in the literature is based upon the location of the ectopic orifice. This system ignores other aspects like the degree of bending (chordee) of the penis, that is descisive for the severity of the malformation. In this type of classification, the hypospadiac case with a subcoronal meatus would always be classified in the group of minor defects. After a proper chordectomy however these cases can have the orifice somewhere in between the penoscrotal area and the subcoronal region, depending on the severity of the chordee.

These problems have been recognized by Culp and Roberts (1968), who introduced a classification of the hypospadias after an adequate chordectomy. This classification is used in Table I, in which the incidence is given of the various types of hypospadias.

Table I: Location of the urethral meatus after
correction of the chordee (Culp and Roberts 1968)

Position of orifice after chordectomy	Incidence
Penile	28.4 %
Penoscrotal	44.8 %
Scrotal	16.5 %
Perineal	10.3 %

Total number of patients: 253

We think, that only a classification, that incorporates the two main features of the hypospadiac entity (the dystopy of the meatus and the chordee) can adequately describe the malformation. For this classification the degree of bending is divided in three categories: mild, moderate and severe. We speak of a mild bending if the distal part of the corpora does not deviate more than 15 degrees from the virtual axis of the more proximal part of the

corpora. In moderate cases the bending should not exceed the 35 degrees. All deviations over 35 degrees are catagorized as severe cases.

Combining these two elements a suitable classification can be obtained, that allows a better description of the anomalies.

Table II: Relationship between the location of the orifice and the presence and degree of chordee in our patients (87 primary cases and 5 patients out of the "cripple" group with reliable preoperative data)

Location of orifice							

Chordee	normal*	coronal	subcoronal	penile	penoscrotal	scrotal	perineal

absent			2	3			
mild		9	16	8	3		
moderate		8	13	4	4	2	
severe	3	2	7	2	4		2
	3(3.3%)	19(20.6%)	38(41.2%)	17(18.5%)	11(11%)	2(2.2%)	2(2.2%)

* The so called hypospadias sine hypospadia
(see chapter I.5 p. 23)

The overall review of larger series shows that in the majority of cases of hypospadias the ectopic orifice is located in the more distal part of the penis (Table III). From these data, however, no information can be given about the degree of chordee of these patients, which illustrates that the severity of the chordee is still not frequently used to classify hypospadias.

Table III: Classification according to the location of the orifice
before corrective surgery
(Total number of patients: 2948)*

	Location of orifice				hypospadias sine hypospadias
	(sub)coronal	penile	scrotal	perineal	
number of					
patients	1308	1784	174	70	12
percentages	44%	43%	9%	2%	< 1%

*Compilation of large series (1974-1985)

(Sweet et al. 1974, Heiss and Helmig 1974, Svensson 1979,
Shima et al. 1979, Johanson and Avellan 1980, Neto et al. 1981,
Marberger and Pauer 1981)

1.3 The morphogenesis

1.3.1 Normal development

The development of the external male genitalia is only possible under the influence of testosterone.

This hormone has to be produced at the proper time in sufficient quantities and it has to be metabolized by specific enzymes at special receptor sites, in order to allow a normal development of the male genitalia (Siebers et al. 1980). If the production of testosterone is blocked, the development of the individual will be in a female direction. He speaks in this context of an induced (male) or of a constitutive (female) sex.

This differentiation in sex, induced by testosterone, involves also sequential, coherent changes in the developing urinary tract (Wolffian or mesonephric duct) and in parts of the intestinal

tract (the cloaca and the cloacal membrane).

Parts of the developing female genital tract (Mullerian duct) have to regress, during this development.

Any disturbance at one of these levels can cause malformations in the final appearance of the male genitalia, varying from a phenotypical female to a phenotypical male with mild hypospadias.

At fertilisation the genetic sex of an individual is determined. This does not mean, that an 46 XY individual will automatically become a phenotypical male. The coherent action of many pairs of genes is necessary to accomplish this outgrowth, starting with the differentiation of the indifferent gonad into a testis.

The Y chromosome, or to be more precise, a gene located on the short arm near its centromere is of critical importance for this gonadal differentiation. It codes for the expression of an antigen (H(istocompatibility) Y antigen, HY antigen, Billingham and Silvers 1960) which is naturally limited to the XY heterogenetic individuals (Wachtel et al. 1976).

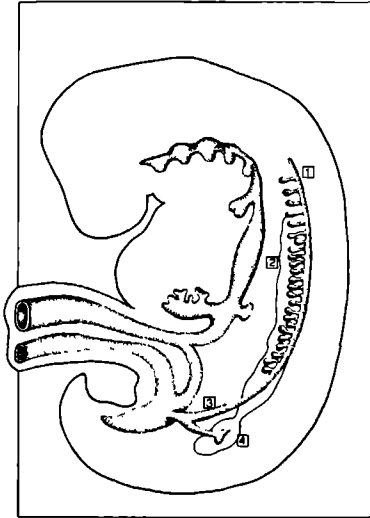
Regulated, not only by this Y chromosome, but also by some autosomes as well, the HY antigen is responsible for the differentiation of the primordial gonadal cells into testicular tissue. It also causes an increase in receptor susceptibility of the cell surface for luteinizing hormone and human chorio-gonadotropic hormone (HCG), that is necessary in the process of gonadal differentiation (Wolf 1980, 1981, Wachtel and Koo 1981, Golimbu 1984).

Disturbances in the chromosomal differentiation of 46 XY or 46 XX are not so rare as previously thought. Investigations of abortion material show an incidence of abnormal development in 25% (Goedde and Hoo 1979).

The development of the urinary tract of which some parts play a role in the genesis of the male genitalia preceeds the gonadal differentiation. In the cranial parts of the embryo (segmental in origin) the nephrogenic ridge is also segmented and every nephrotome forms a vesicle, that is interconnected to the next (more caudally located) segment by a tube: the pronephric duct. All these vesicles desintegrate after the next one has formed. Only the pronephric duct survives in the more caudally developing

structures (only partially segmented), as the mesonephric or Wolffian duct. This Wolffian duct grows out more caudally in the complete unsegmented area of the embryo and makes contact at both sides with the cloaca at a paramedian point. It induces in the 4th week of gestation the forming of the primitive glomeruli of the mesonephros, of which the collecting systems make contact with this Wolffian duct. In the same week the ureteric bud appears at the dorso-medial aspect of the Wolffian duct near its entrance in the cloaca. This bud grows out more cranially and makes contact with the totally unsegmented meta-nephrogenic blastema (just caudally of the mesonephric zone) and induces the formation of the permanent kidney (fig.5).

Rudimentary pronephros [1]
= forekidney
present from third-fourth week



Excretory tubules of the
mesonephros = midkidney [2]
differentiates in the fourth
week, regresses in the eighth
week

Mesonephric duct [3]
(Wolffian)

The permanent kidney [4]
metanephros = hindkidney
begins to differentiate in
the 5th week

Fig. 5: Sequential development of the pronephros,
mesonephros and metanephros in a human embryo

In this period (end of the fourth week) the primitive gut ends at the ventral aspect of the embryo in the cloaca, separated from the amniotic fluid by the cloacal membrane, that exists out of only two layers of cells: ento- and ectoderm.

The cloaca does not serve for long as a common outlet for the urogenital and digestive systems, but is soon separated by an ingrowth of ento-dermal mesenchyme (the urorectal septum) starting cranially of the allantois. This ingrowth takes place in cranial to caudal and lateral to medial direction. This mesenchyme divides the cloaca in an anterior urogenital sinus, and a posterior rectal canal (6th and 7th week) (fig. 6).

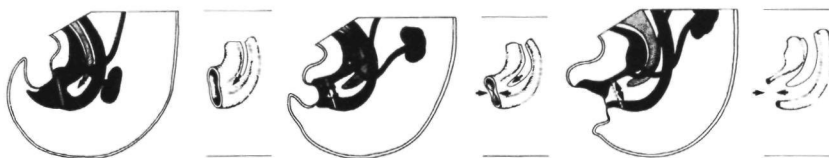


Fig. 6: Division of the cloaca and formation of the urogenital sinus and the anal canal (after Stephens 1983)

During the development of the Wolffian duct, the coelomic epithelium in this area indentates and forms a new tube: the Müllerian duct. Its development follows closely the caudal movement of the Wolffian duct. At the level of the caudal segment of the mesonephros it crosses from a lateral position to a medial one in respect of the Wolffian duct. The Müllerian duct and the Wolffian duct then fuse in the midline before entering the urogenital sinus. This crossing point is important in the further differentiation of both ducts. The desintegration of the Müllerian duct in the male (Jost, 1953, 1958) starts at this point, gradually extending, but mostly never reaching the most caudal and cranial parts, which can remain in the male as

respectively the utriculus and the appendix testis (Jirasek 1977). The gonadal ridge caudally to this crossing point develops as gubernaculum Hunteri and forms the connection between the testis and the scrotal wall (fig. 7).

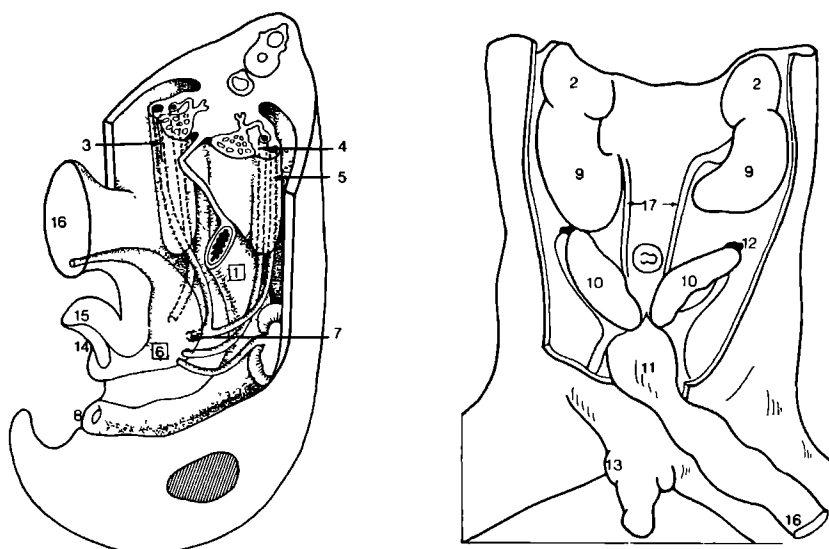


Fig. 7: Wolffian and Müllerian duct (completely developed 7th week)

1. digestive tract
2. adrenal gland
3. nephrogenic ridge
4. Wolffian duct
5. Müllerian duct
6. urogenital sinus
7. utriculus
8. anal pit
9. kidney

Müllerian duct desintegrated 9th week (after England 1983, Langman 1985)

10. testis
11. bladder
12. Müllerian duct remnants
13. labio-scrotal swellings
14. uro-genital folds
15. genital tubercle
16. umbilical cord
17. ureter

During the development of the mesonephros the primordial germcells migrate from their position in the dorsal mesentery of the primitive gut to their definite position medially of the

Wolffian duct. These cells are then located in the so called genital ridge. Subsequently the coelomic epithelium proliferates into primitive sexcords around those cells. In the male these cords will differentiate into primitive seminiferous tubules, which make contact with the mesonephric tubules at the end of the 6th week, establishing the first urogenital connection.

The differentiation of the cranial parts of the Wolffian duct into the rete testis (partially), caput, corpus and cauda epididymis, is influenced by testosterone, produced by the primitive testis, and locally active in the surrounding tissues (around the 7th week).

At that moment developing Sertoli cells start the production of a non-steroidal substance, without androgen properties: the so-called Muller Inhibiting Factor (Jost 1953, 1972).

It is produced independently of the production of human gonadotropine, of either maternal or fetal origin. A high level of the Muller Inhibiting Factor is maintained throughout the entire gestational period. It causes the ipsi-lateral desintegration of the Mullerian duct. Any disturbance in the production of this hormone, either unilaterally or bilaterally, blocks the normal sequence of developmental events and leads to persistence of Mullerian (female) structures in even chromosomal normal males.

The Leydig cells are differentiating in the 8th week out of the mesenchyme between the testicular sexcords. This is caused by the rising levels of maternal human choriongonadotropine. From the 12th week on, the fetal pituitary, producing the luteinizing hormone (LH), stimulates the further development of the Leydig cells. During the 12th-16th week these cells reach their maximum both in number and level of metabolism, providing the highest testosterone levels in the prenatal period, that coincides with the development of the external genitalia. The formation of the external genitalia depends on the presence of this testosterone.

In the 6th week the lateral proliferations around the urogenital membrane fuse ventrally in the midline and form the genital tubercle. The mesoderm around the anal membrane proliferates also and forms the anal tubercles (fig. 8).

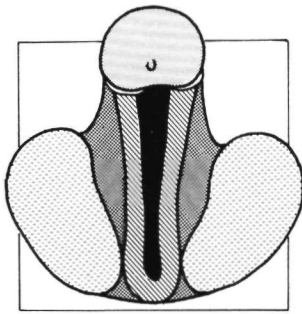


Fig. 8: Development of the genital tubercle, the labioscrotal swellings and the anal tubercles

This mesoderm has the capacity to convert testosterone to dihydrotestosterone in the male by the enzyme 5-alpha-reductase. This is one of the first signs of sexual differentiation.

The genital tubercle starts to grow rapidly from the moment that testosterone is circulating and in about the 10th week, the glans is recognizable as a well marked formation. The anal tubercles take part in the formation of the perineum and the ventral wall of the anal canal. Subsequently the genital folds migrate medially and form the perineal raphe (Fig. 9). At this stage both the urogenital sinus and the anal canal are open.

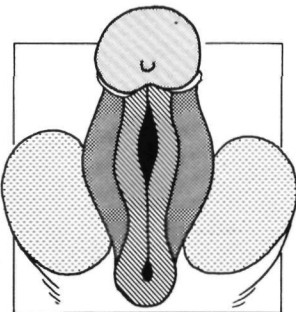


Fig. 9: Development of the genital tubercle into a penis and formation of the perineal raphe

During the proliferation of the genital tubercle the entodermal

skin layers form the urethral plate at the ventral aspect as a continuation of the urogenital sinus. This urethral plate gradually deepens and forms then the urethral groove lined by the urethral folds. The urethra proximal of the outgrowing penis is at that moment already completed. In the 12th week the urethral folds start to close in the most proximal part of the outgrowing genital tubercle forming the bulbous urethra (fig. 10).

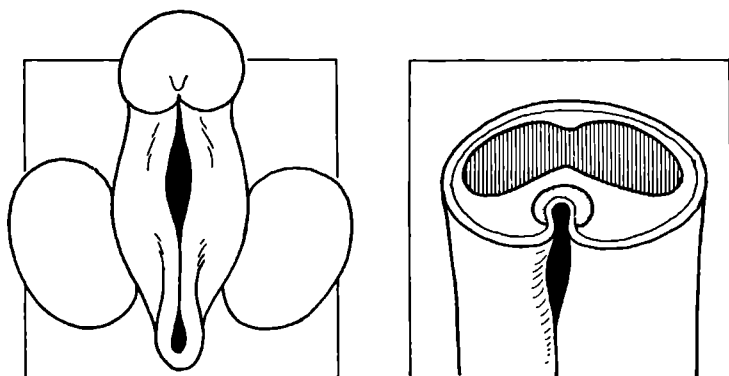


Fig. 10: Closing process of the urethra

The proliferating mesenchyme starts to surround the urethra and separates it from the ectodermal skin layer, including the bulbo-urethral musculature (fig.11).

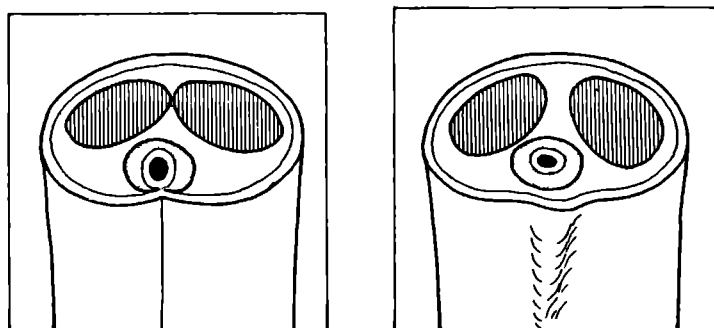


Fig. 11: Mesenchymal proliferation and formation of the superficial subcutaneous layers, the spongiosal and cavernosal corpora

As result of this extending closing process, the urogenital orifice migrates towards the glans. The surrounding mesenchyme forms in this penile part the spongiosal corpus and the fascial layers. In the dorsal parts of the genital tubercle this mesenchyme condensates, and forms bilaterally the cavernosal corpora, the nerves and bloodvessels.

This closing process is completed till the coronal sulcus is formed at approximately the 14th week. At this level the orifice stays open for a longer period of time and the urethral groove is deeper than normally (fig. 12).

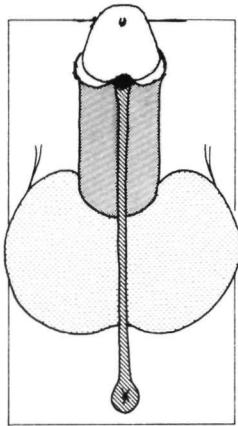


Fig. 12: Completed urethral closure up to the coronal sulcus

At the coronal level the epithelium starts to proliferate in the 12th week at each side of this urethral opening and gradually grows around the glans in the following eight weeks. During this outgrowth the glans closes in the midline and both edges of the unrolling skin fuse, forming the frenulum. In the outgrowing glans, the urethral groove deepens and fuses in a later phase with the ingrowing ectodermal tag at the top of the glans.

This ectodermal proliferation is formed from about the end of the 10th week. Under the influence of testosterone it starts to grow into the center of the glans in proximal direction to the level of the coronal sulcus. After canalization the fusion takes place (Glenister 1954, Stephens 1983) (fig. 13).

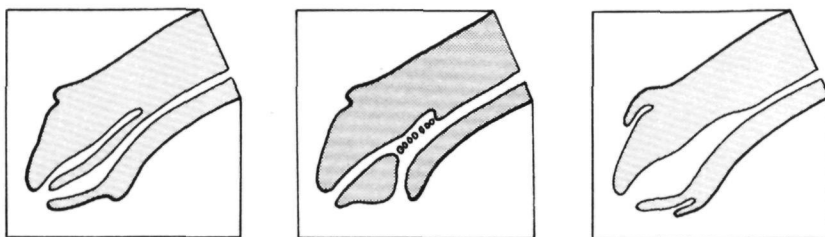


Fig. 13: Ingrowth and canalization of the epithelial tag forming the navicular fossa

At the base of the outgrowing genital tubercle in the 8th week a sharp delimitation appears (phallic groove), separating the future penis from the outgrowing labioscrotal swellings. These swellings migrate posteriorly during their outgrowth and lengthen in this way the raphe. If this movement is interrupted the penis remains dystopic (the engulfed penis) and because of a shortness of skin at the ventral side a bending of the penis is caused (fig. 4).

At the 9th month the descend of the testis completes the appearance of the external male genitalia.

1.3.2 The ontogenesis of the hypospadiac entity

Hypospadias is the consequence of not totally completed virilization process.

A conditio sine qua non is the presence of a maximal production of testosterone by the Leydig-cells during the short time in which the development of the external genitalia takes place. If this production maximum is too early or the involution of the cells takes place in a period the external genitalia are still developing, the closing process of the urethral groove will be interrupted and a ventrally located urethral orifice is the result.

Distally of this ectopic orifice the urethral groove is still in its place, extending up to the level of the coronal sulcus. The glans will not close around the glandular urethral groove and

will keep a rather flattened appearance with a ventral tilt. The proximal mesenchyme is not able to fuse in the midline. The consequence is that differentiation of the mesenchyme into Bucks' fascia, the tunica dartos and the spongiosal tissue is disturbed in this chronological order (Kaplan and Brock 1981). This tissue is atretic, forming fibrous bands, which usually extends in a fan-shaped area, tethering the distal penile shaft. This causes a bending of the penis in erection (Devine 1983)(fig.14).



Fig. 14: Bending of the penis, due to chordee

Normally, the potency of the mesenchyme to differentiate does not stop abruptly at the level at which the urethral groove stops closing. It is possible to find virtually normal spongiosal tissue, distally of the orifice, together with the more fibrotic elements of the primordial fascia (fig. 15).

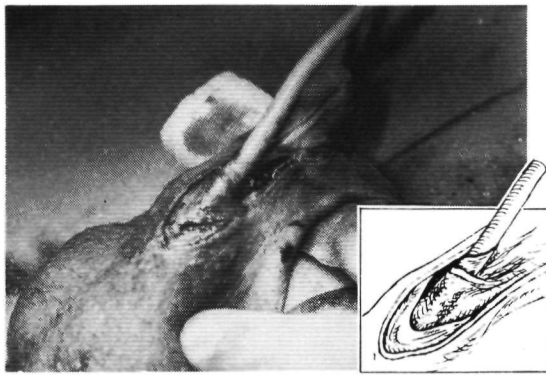


Fig. 15: V shaped form of both spongiosal and urethral plate remnants

We agree with Devine (1973, 1983), that both, urethral plate remnants and more or less fibrotic spongiosal and fascial primordial tissue form the base of the chordee, which is in line with the early descriptions of Theile (1847).

The fact that the urethral groove does not close, also prevents the fusion of the skin layer in the midline. So a frenulum can not be formed. The skin edges will be located at each side of the urethral groove, creating the V shaped ventral skin defect and forming a dorsal hood of skin. They end then in the so called "dogears", which in our opinion form the epithelial proliferation site of the frenulum in normal circumstances (fig.3).

1.4 Anomalies associated with hypospadias

1.4.1 Cryptorchism

The most frequently associated anomaly in hypospadias is the incomplete descend of the testis. The overall incidence of maldescensus testis (in the English and American literature referred to as Cryptorchism) in the general male population at about one year of age is considered to be 1% (MacKellar et al. 1984).

In hypospadiac cases the incidence of maldescensus testis is much higher, it varies in literature from 2.2 to 19.3% (Table IV).

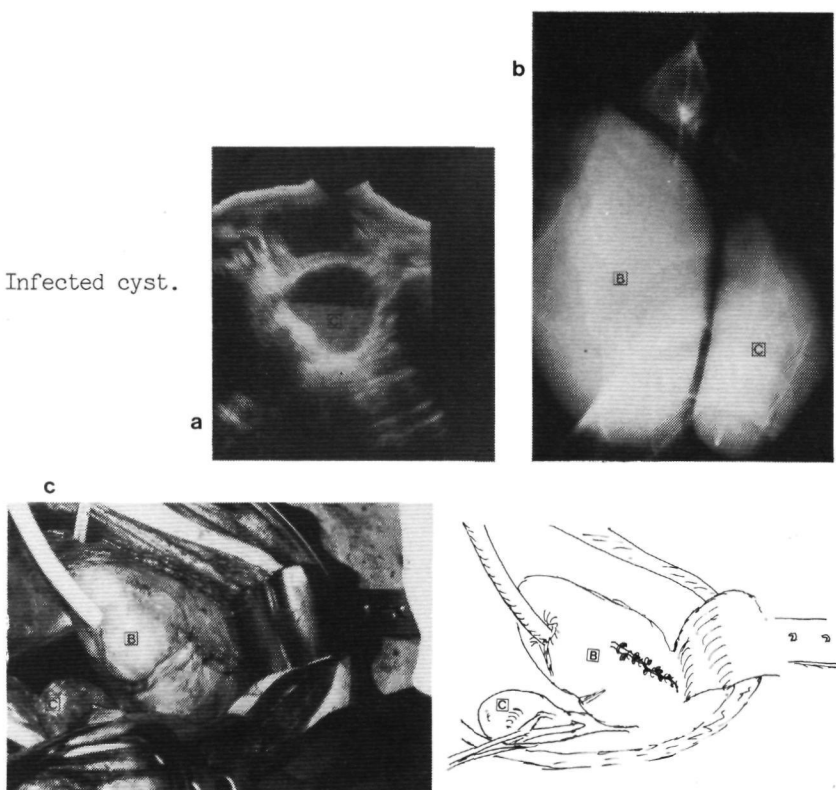
Table IV: Percentage of maldescensus testis
(Cryptorchism) in hypospadias

Author		number of patients	percentage of cryptorchism
Sørensen	(1953)	274	19.3
Kennedy	(1961)	489	10
Culp and Roberts	(1968)	422	11
Heiss and Helmig	(1974)	813	13.9
Svensson	(1979)	625	6
Bauer et al.	(1981)	177	5
Khuri et al.	(1981)	1076	9.3
Källen and Winberg	(1982)	1357	2.2
Leung et al.	(1985)	1314	2.4

1.4.2 Utriculus

The persistence of this structure in hypospadias is well known and already adequately described by such early anatomists as Morgagni (1761) and Theile (1847). The incidence of this anomaly differs considerably in literature. Mostly it is only described in severe cases (Campbell 1951, Smith and Forsythe 1959, Williams 1982). Howard (1948) found it in 6 penoscrotal cases and in 2 perineal cases, out of 14 cases, while Devine et al. (1980) found an overall incidence of 14% in a group of 44 moderate to severe hypospadiac cases. Ikoma et al. (1985) reported an incidence of 27.5% in a group of 280 radiologically investigated hypospadiac cases. In our own material we found it in 2 out of 115 cases. In both cases the utriculus cyst became symptomatic, causing recurrent urinary tract infections. Fig. 16 illustrates one of these cases.

Infected cyst.



Intra operative view

Fig. 16: Utricle cyst Grade IV in a 9 year old boy.

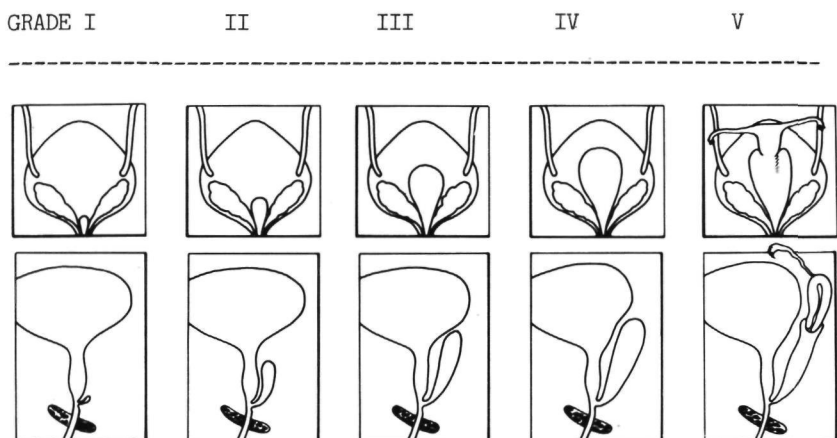
a. Sonographic view of the infected cyst.

b. Genitography combined with a cystogram.

c. Intra operative view

B = Urinary bladder; C = utriculus cyst

In order to standardize the classification of utriculus cysts, we propose the following grading system based upon the preoperative urethrocystoscopy (fig. 17).



- I The cyst is just a few millimeters in depth
 II The cyst extends till the bladder neck
 III The cyst extends beyond the bladder neck
 IV The cyst extends beyond the seminal vesicles
 V The cyst is combined with other remnants of the Mullerian duct

Fig. 17: Proposed classification of the utriculus cysts

Since 1983 we scrutinize every patient with hypospadias for the presence of an utriculus cyst. In 13 out of 25 consecutive hypospadiac patients we found an utriculus cyst:

Grade I in six cases, grade II in four cases, grade III in three cases, and no grade IV or V.

This is an incidence of approximately 50%, although severe cases were overrepresented in our series.

1.5 Hypospadias sine hypospadias

Another malformation, however not belonging to the hypospadiac entity (the orifice is not dystopic), is the Hypospadias sine hypospadias.

It is a rather rare anomaly. Culp and Roberts (1968) saw it in 14 out of 422 cases. Khuri et al. (1981) mentioned it in 13 out of 1076 cases. In our series we found it in 4 out of 115 cases.

Several of our cases however did raise serious doubt as to whether this entity is so much different from those cases who have an ectopic orifice.

In many cases we think that there is a secondary stimulus that just enables the urethra to close.

This "secondary repair" pattern results in a penis with a normally located meatus: the ingrowing epithelial tag was able to contact with the deepening urethral groove, that canalizes by secondary fusion patterns. In these patients as in patients with an ectopic orifice, every separate layer can be insufficient, while others are completely differentiated. These differentiation disorders resulting in the hypospadias sine hypospadias can be categorized into five groups (Devine and Horton 1973, 1977, Devine 1983).

In the first type the tunica dartos and partially the Bucks' fascia are insufficient. Chordee can be found both ventrally and dorsally of the normal spongiosal tissue.

In the second type neither the tunica dartos nor Bucks' fascia and the spongiosal corpora are formed. Severe chordee will be found around that part of the urethra, that only exists of two cell-layers: ecto- and entoderm.

In type three only the differentiation of the tunica dartos is incomplete and the chordee can be found laterally of the normal urethra.

In type four only the skin layer is insufficient and the chordee is only a cutaneous one, that can be cured by a simple skin rearrangement.

In the fifth type all layers are macroscopically normal but a hypoplasia of the ventral part of the cavernosal corpora is found.

This condition is sometimes encountered in different degrees of hypospadias (Voillemier 1964).

Table V: Hypospadias sine hypospadias

Disturbances in the development of the different tissue layers causing chordee in cases with a normally located meatus

Type	Skin	Tunica Dartos	Bucks' Fascia	Spongiosal Corpora	Urethra	Cavernosal Corpora
I	normal	not developed	deficient	normal	normal	normal
II	normal	not developed	not developed	not developed	2 cell layers	normal or ventrally hypoplastic
III	normal	deficient	normal	normal	normal	normal
IV	deficient	normal	normal	normal	normal	normal
V	normal	normal	normal	normal	normal	hypoplastic ventrally

After Devine (1983).

1.6 Epidemiology

The incidence of the hypospadiac entity ranges in literature from 0.076% (Neto et al. 1981) to nearly 0.8% (Sweet et al. 1974).

An overview of the incidence reported in the literature is given in Table VI.

Table VI: Incidence of Hypospadias

Reference		Number of live birth		Incidence per 1000 deliveries	
		male	all	male	all
Böök	1951	22.180		0.8	
Wallace et al.	1953	81.377		1.2	
Sørensen	1953	27.613		3.3	
Roberts and Lloyds	1973		92.982		1.1
Sweet et al.	1974	12.776		8.2	
Avellán	1975		480.607		1.39
Neto et al.	1981		423.839		0.76
Källén and Winberg	1982		706.532		1.92
Matlai and Beral	1985			1.5-3.6	
Leung et al.	1985	295.656		4.44	

Probably this great difference in the incidence of hypospadias is caused by differences in selection of patients.

One of the first studies was performed by Rennes (1831), who found ten hypospadiac cases among 3000 recruits. Another study was Campbell's (1951) post mortem series on 12.280 boys, in which he found an incidence of 0.09%. Sørensen (1953) published a very careful study concerning the hypospadias incidence in Denmark. He found in 27.613 live male births ninety hypospadiac cases. An incidence of 0.33%. This incidence figure remained for a long time the index number quoted in the literature.

In recent years in several countries an increase of the incidence was found. In Sweden an increase from 0.08 to 0.13% was noted by Källén and Winberg (1982). In Hungary Czeizel (1985) found an increase from 0.12 to 0.47%. In Wales and England an increase from 0.16 to 0.36% is reported by Matlai and Beral (1985). An explanation for this increase in incidence has not yet been found. We are not able to give an exact figure for our country, because

registration of this malformation is not obligatory and furthermore the registration is hampered by the fact that half of the deliveries take place at home. It seems reasonable however, that the incidence in our country may be estimated at a level in between the two extremes. In general an incidence of one hypospadiac boy in every 300 male births is accepted.

1.7 Aetiological considerations

1.7.1 Exogenous factors

Throughout the literature an exogenous factor causing hypospadias, has frequently been advocated. Some investigators found evidence for an exogenous factor in experimental animal studies since the administration of sexhormones to the mother resulted in changes in the external genitalia of the male offspring (Moore 1944, Dantchakoff 1953).

Recently epidemiological studies of Czeizel (1985) revealed an increase of hypospadias in children of mothers with a very high intake of sexhormones. Since hypospadias is more frequently seen in older mothers, several authors relate this to level of human choriongonadotropines (Loughran 1948, Büchi 1950, Aarskog 1979, Neto et al. 1981, Angerpointer 1984 and Czeizel 1985), although others were not able to confirm this observation (Sweet et al. 1974, Avellan 1977, Källen and Winberg 1982).

That viral infections (Albaugh 1945) or vitamin deficiencies (Warkany 1948) were responsible for the development of hypospadias could not withstand critical analysis (Sørensen 1953).

1.7.2 Heredity

This is the oldest epidemiological feature, that has been recognised in the hypospadiac cases (Kopp 1810). From the studies of Sørensen (1953) and Bauer et al. (1981) it has become clear, that

a single gene heredity theory cannot be responsible for the hypospadiac entity, but that a multi-factorial mode of inheritance must be accepted. Recently a possible homozygosity for recessive genes has been described as a possible cause in some families (Frydman et al. 1985).

The incidence of hypospadias among relatives from a hypospadiac patient is higher than the overall incidence in the population, although the incidence of affected relatives varies considerably in the literature. This is probably due to a selectional bias, since the incidence of affected relatives is higher in more severe cases of hypospadias, (Sweet et al. 1974, Neto et al. 1981). Table VII.

Table VII: Incidence of familial occurrence of hypospadias

Reference	number of families studied		number of families with one or more relatives with hypospadias	percentages of cases with at least one affected relative
Sörensen	1953	103	29	28.1%
Sweet	1974	107	9	8 %
et al.				
Neto	1981	309	19	6.1%
et al.				
Bauer	1981	307	64	21 %
et al.				
Angerpointer	1984	458	20	6.8%
Leung et al.	1985	1314	42	5.4%

1.7.3 Modern concepts on the aetiology of hypospadias

In recent years it has become evident that besides chromosomal aberrations (Goldschmidt 1931, Howard 1948, Campbell 1951) also hormonal disturbances during fetal development can cause

hypospadias.

We now accept two forms of pathways, resulting in the hypospadiac entity.

1 The directly inheritable single-gene disorders. This group can be divided into two subgroups:

- a chromosomal aberrations like Klinefelter, Turner, Down, Prader-Willi and several other syndromes (Petrykowski 1981, Rosenbaum 1985).
- b monogene inheritance like the Leopard, Rieger and several other syndromes (Popelier 1981).

A discussion about these relatively rare anomalies is beyond the scope of this study.

2 The multi-factorial model with non directly inheritable disorders.

In this group we have to distinguish between the familial and the non-familial cases.

The familial cases can be explained by an autosomal, recessive inheritance with low penetrance. Several factors have to cooperate to overcome the threshold of the penetrance of one or more genes (Sørensen 1953). The non-familial cases are characterized by a multifactorial-polygenic cause, without any recognizable pattern of genetic inheritance (Keenan 1980, Glatzl 1981, Berg 1983).

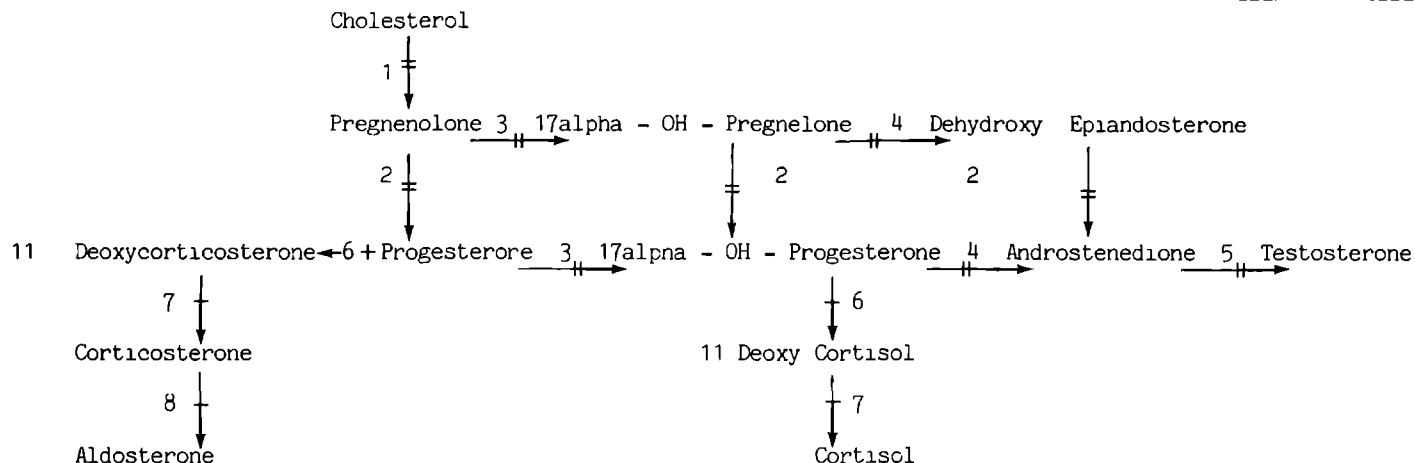
An attractive theory was proposed by Kallen and Winberg (1982), who were looking for an explanation of the increase in incidence of the hypospadiac entity. They proposed the presence of a gene which is responsible for reduced testicular function, both in the embryo as in the adult: the subfertility theory.

Krause et al. (1984) speak in this context of an intrinsic testicular factor. This would explain the decreased production of local and hormonal testosterone and the possible subsequent decrease in production of the Müller Inhibiting Substance. The

latter can be responsible for the Mullerian remnants in hypospadiac cases. This would also explain the high incidence of testicular abnormalities in fathers of hypospadiac boys (Sweet et al. 1974) and the high incidence of maldescensus testis in hypospadias. The various expressions of this entity, might be the consequence of a varying expression of this gene, due to different threshold factors. This would emphasise the fact, that the more serious forms of hypospadias are more frequently associated with other concomittant anomalies (Svensson 1979, Khuri et al. 1981). Other disturbances can lead to an imbalance of the hormonal production. The first hormone, that can be disturbed either in quantity or in time of production is HCG of placental and later pituitary fetal origin. Without this hormone the testosterone production is impossible with all the consequences for the formation of the male genitalia.

Some disturbances in the production of adrenal steroids can cause secondary changes in development of the outer genitalia. Table VIII.

Table VIII: Possible enzymedefects in the testosterone production causing incomplete masculinization in male individuals or secondary virilization in female individuals



—||—> Enzyme Blocks causing incomplete masculinization
 —> Enzyme Blocks causing virilisation in females by blocking the formation of cortisol: build up precursors have androgen activity!

1 : 20alpha	Hydroxylase, 22alpha Hydroxylase, 20, 22 Desmolase	5 : 17	Ketosteroid reductase
2 : 3beta	Steroiddehydrogenase	6 : 21	Hydroxylase
3 : 17alpha	Hydroxylase	7 : 11	Hydroxylase
4 : 17.20	Desmolase	8 : 18	Hydroxylase

Adapted from Petrykowski 1981, Glatzl 1984, Allen 1985.

In the female individuals this can cause secondary virilization (fig. 18), in male individuals this can cause insufficient masculinization (fig. 19).

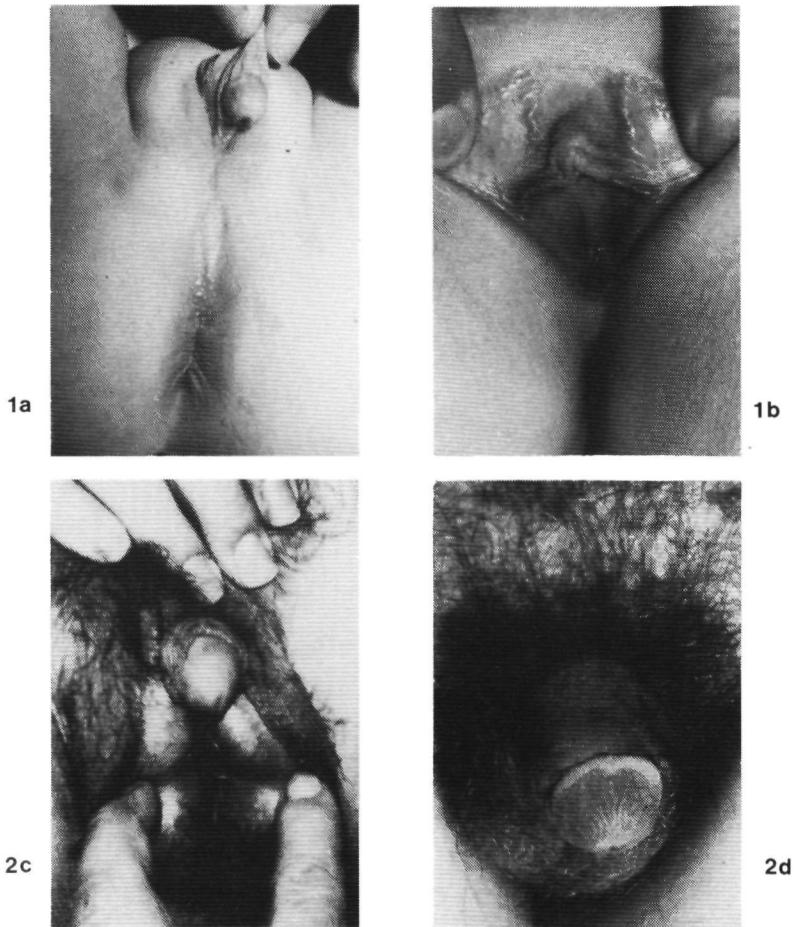
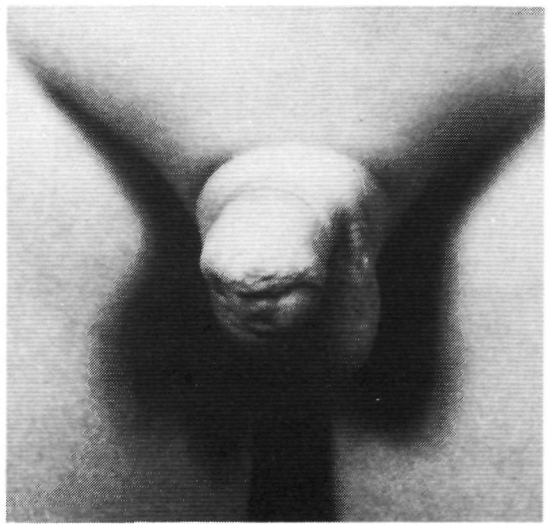


Fig. 18.1: Adreno-genital syndrome of a 5 year old XX female corrected in female direction
a. before correction (5 years of age)
b. after correction (8 years of age)
.2: Adreno-genital syndrome 14 year old XX individual after gonadectomy and hormonal suppletion, corrected in male direction
c. before correction
d. after correction (testicular prostheses in situ)



a



b

Fig. 19: a. 2 year old boy with a 20-22 Desmolase Insufficiency
before operation
b. Idem 9 month after operation

Besides a normal hormonal production, also an appropriate response of the target tissue for these hormones is necessary for a normal development of the male genitalia. There are indications, that in certain hypospadiac cases this target tissue can have insufficient receptor function for these hormones as a possible cause for the malformation (Jost 1972, Coulam et al. 1983, Jukier et al. 1984).


If so, the outgrowth of the sensitive tissues (the genital part of the urogenital sinus) is interrupted (fig. 20).


UROGENITAL SINUS
DIHYDROTESTOSTERONE DEPENDANT

WOLFFIAN DUCT
TESTOSTERONE DEPENDANT

		Caput (partially)
Prostate	Epididymis	Corpus
		Cauda
Urethra distal of the verumontanum	Ductus deferens	
Penis	Seminal vesicle	
Scrotum		

urogenital sinus 

Wolffian duct 

Mullerian duct 

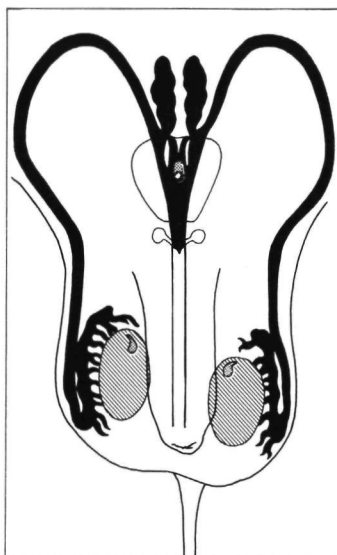


Fig. 20: Hormonal influences on the development of male genital tract (after Siiteri and Wilson 1974).

The role of hormonal imbalance in the aetiology of hypospadias is further given by the fact that in hypospadiac patients hormonal stimulation tests lead to lower responses of the testosterone compared to matched control groups (Shima et al. 1981, 1986, Knorr 1985). The consequences of this type of

disturbances are serious, leading to various developmental abnormalities which can range from phenotypically female to severe hypospadias (Table IX).

Table IX: Some enzyme and receptor defects and/or insufficiencies with the clinical consequences.

	Effect on hormonal production and/or availability	Fetal development	Phenotypical aspects after puberty	
			Penis	Breast
1. Enzym or receptor defects causing early hormonal disturbances (HCG or LH insufficiency)	no testosterone production	female	no growth	no or moderate growth
2. Enzymedefects in the testosterone production (Table VIII)	insufficient testosterone production	female to hypospadias	relative growth to small size	no growth till moderate
3. 5alpha-reductase defect and/or insufficiency	normal testosterone production	severe hypospadias	moderate growth	no growth
4. Androgen receptor insensibility	elevated testosterone levels	female to severe hypospadias	hardly any till almost normal growth	hardly any till almost normal growth

Adapted from Keenan (1980) and Glatzl (1981, 1984).

Review of the operative procedures for correction of hypospadias.

2.1 Introduction

In this chapter we will review the various methods that has been used or still are used for the correction of hypospadias. First we will describe the various basic methods that enabled the introduction of corrective surgery for hypospadias. Subsequently we will discuss the various two-stage procedures, in which in general the first session is used for the correction of the chordee and in the one or more subsequent sessions the neo-urethra is formed. Finally the more recently used one stage procedures are reviewed. The method that we have used in our study (the inner preputial island flap technique) will be described in detail in chapter III.

2.2 Basic methods

Since the first description by Galen (131-201 A.D.) in the second century who corrected hypospadias by a canalization technique, little progress has been made in the surgical repair of hypospadias in subsequent centuries. The improvement of closure techniques in the repair of fistulas in the last century, (Cooper 1820, Dieffenbach 1836, 1845) made surgical reconstruction in hypospadias feasible (fig. 1, 2).

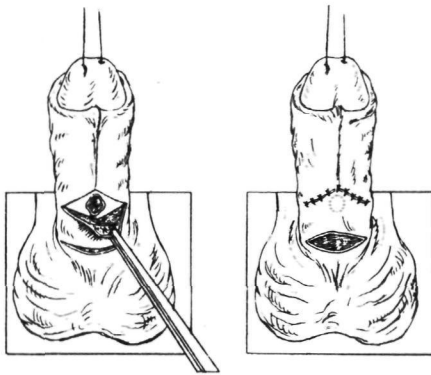


Fig. 1: The use of skin transposition that made reliable fistula closure possible. After Dieffenbach (1836)

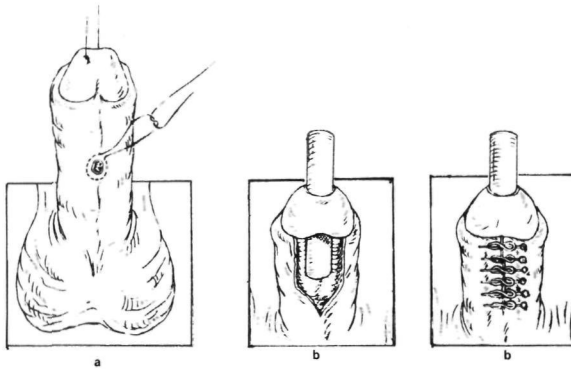


Fig. 2: New closure techniques in fistula repair

- a. the use of the purse string suture
- b. the use of the "needle-suture"

After Dieffenbach (1845)

The operative technique, introduced by Thiersch (1869), for the correction of epispadias, formed a major progress in corrective urogenital surgery (fig. 3).

- a,b: Forming a tube by shifting two lateral based flaps to the contralateral side in an upside down way of the most centrally located flap. The outer flap is then imbricately fixed.
- c,d: Closing the distal "fistula" by splitting the prepuce transversely and delivering the glans through this "buttonhole". The two layers are separately sutured to the edges of the circumferentially dissected "fistula".
- e,f: Closing of the proximal "fistula" by shifting two vascularized flaps, the most central one in an upside down manner.

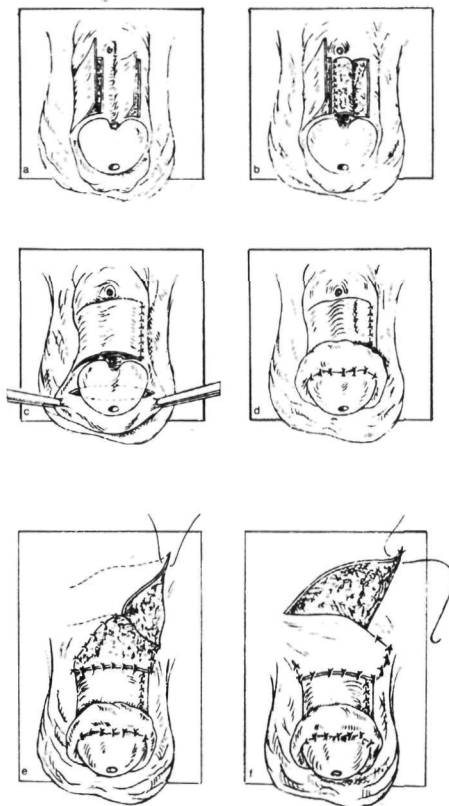


Fig. 3: Principle of skin flap rotation in epispadias repair.
After Thiersch (1869).

These surgical solid principles were readily spread among the European and especially the French surgeons. Based upon this technique basic methods for hypospadias repair could be developed.

2.2.1 The chordectomy

Generally the correction of the bending of the penis (chordee) was considered to be less important than the construction of the neo-urethra.

Mettauer (1842) reported a type of chordectomy much in concordance with the description of Ambroise Paré (1585). He made several transverse cuts in the corpora at the level of the most severe bending. Subsequently these transverse cuts were closed longitudinally with separate inverting sutures.

The type of chordectomy described by Bouisson (1861) became more or less the standard method, in the following years in the reconstruction of hypospadias. He removed all the fibrotic tissue that could be found distally of the meatal orifice (fig. 4).

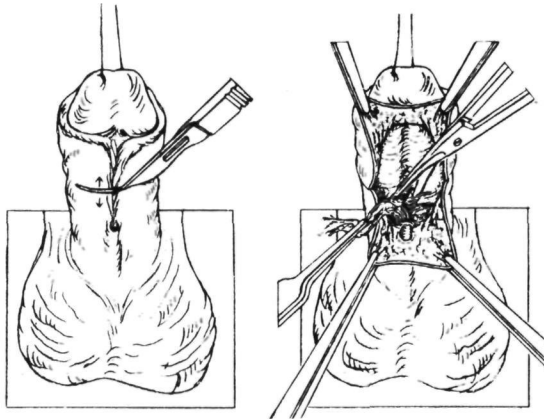


Fig. 4: Chordectomy

Removal of all constricting fibrotic tissue distally from the ectopic urethral orifice.

After Bouisson (1861).

2.2.2 Principle of the technique of imbricated skin flaps

with a tubed or buried epithelial skin strip

The Thiersch technique for correction of hypospadias was introduced by Anger (1874). He used a skin flap rotation at the ventral site of the penis, he could obtain good reproducible results (fig. 5).

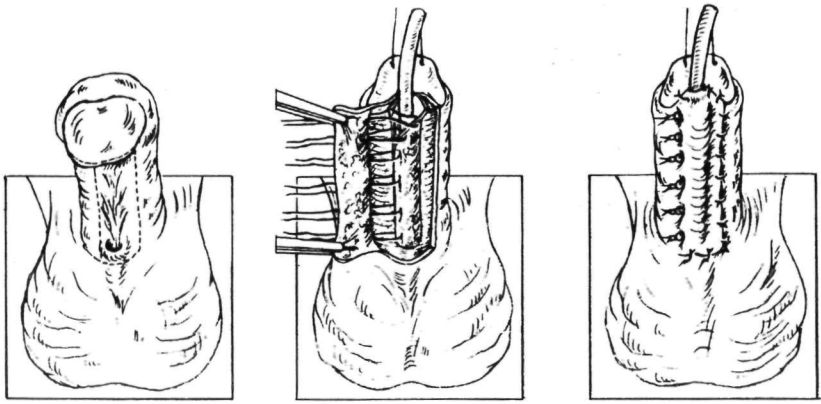


Fig. 5: Hypospadias repair:

Neo-urethroplasty using the Thiersch technique of imbricated flaps. After Anger (1874).

At the same time Duplay (1874) introduced a different approach. He used a ventrally localized strip of epithelium extending from the ectopic orifice to the coronal sulcus (fig.6). In the beginning this strip was always tubularized (Duplay I) but soon a variation was used, in which the tube was not closed (Duplay II) in order to prevent fistula formation. This technique however made a third session necessary in which the ectopic orifice was connected with the neo-urethra. Later on this procedure was incorporated in the second stage in which the neo-urethra was constructed.

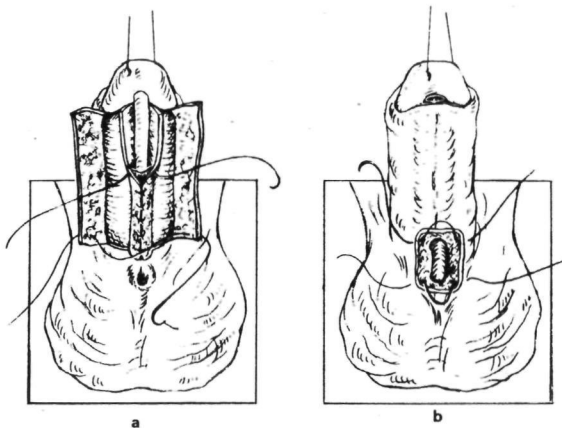


Fig. 6: Hypospadias repair:

- a. formation of the neo-urethra by a tubularized (Duplay I) or a non-tubularized (Duplay II) strip of skin (second stage)
- b. the third session (after chordectomy and the urethroplasty): after preparing a single flap of skin, in which both orifices were localized, the undermined skin edges are closed watertight. After Duplay (1874).

2.2.3 Principle of the meatal-based flap with ventral

cover by prepuce

Wood (1875) working at King's College Hospital in London was the first who described the use of the meatal-based flap for the urethral neoplasty, in combination with the Thiersch (1869) type of preputial cover.

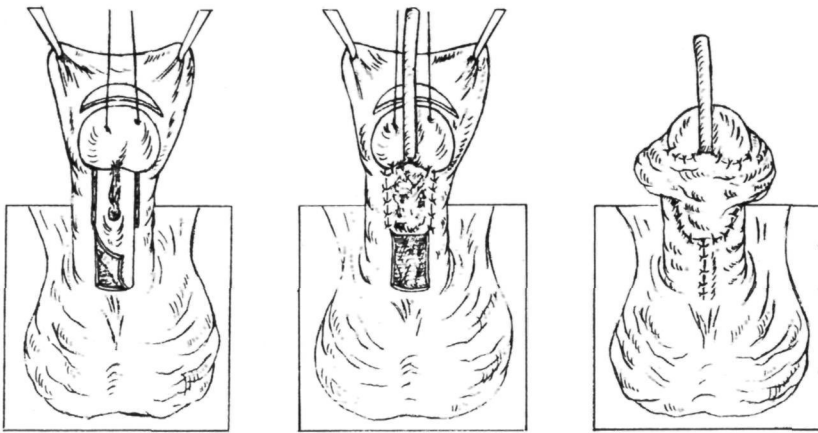


Fig. 7: Hypospadias repair: using a meatal-based flap and preputial cover, bringing the prepuce from the ventral to the dorsal aspect of the penis through a buttonhole. After Wood (1875).

The use of prepuce for compensation of the shortness of ventral penile skin was popularized by one of his successors Edmunds (1913). He chose a planned three stage technique knowing that the majority of the contemporary used methods usually needed more than two operations to give an acceptable result. The chordeectomy was a two-stage procedure starting with the formation of a preputial tube flap. The second stage was used to remove the chordee and to cover the ventral aspect using the enrolled preputial tube (fig. 8).

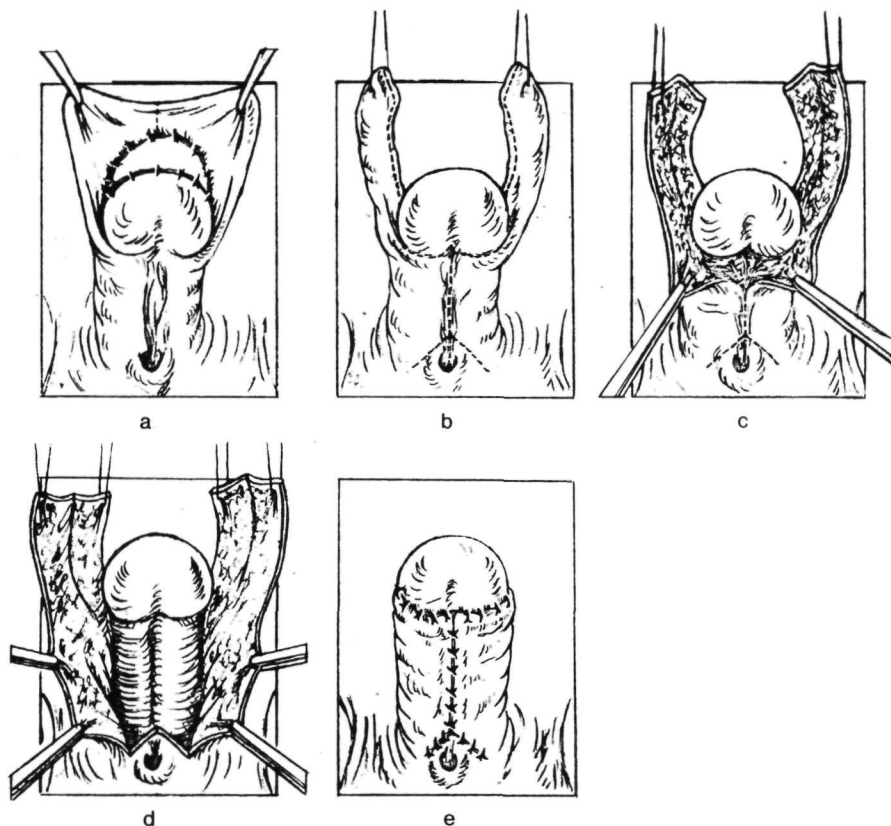


Fig. 8: Hypospadias repair: use of the prepuce as compensation for dorsal skin shortness after chordectomy

- a. first stage used to form a pedicled double flap from the prepuce
- b,c,d. second stage used to cover the ventral surface after chordectomy with the enrolled preputial tubes. In the first stage the neo-urethroplasty was performed.
- After Edmunds (1913).

The necrosis of the flap was avoided by this staged procedure. The urethra was then reconstructed in the third stage using one of the Duplay (1874) methods.

Ombredanne (1911) introduced another technique making use of the redundant prepuce at the dorsal penile side. He reverted to the purse string suture technique to build up a distal urethra without the serious problems of fistula formation as seen with

Duplay's type of repair. He used the buttonhole technique to transfer the dorsal skin to the ventral side (fig. 9). This combined method, with minor variations, has been used until recently (Matthieu 1932, Heybroek 1964, Barcat 1973).

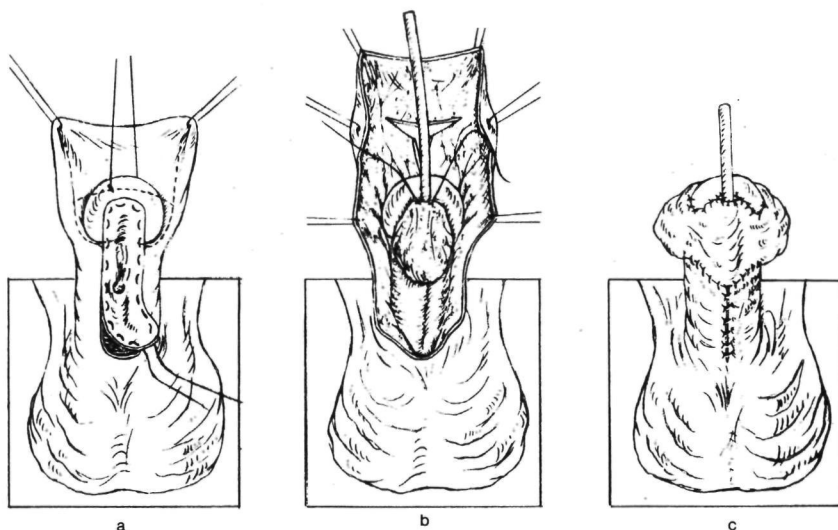


Fig. 9: Hypospadias repair:

- a. combining the purse string suture technique for the formation of a meatal-based flap
- b. preparing the prepuce by dissecting the two layers
- c. after transposition of the prepuce to the ventral aspect through a buttonhole, it is used as a replacement for the ventral penile skin.

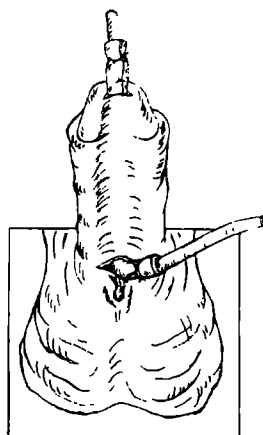
After Ombredanne (1911).

2.2.4 Principle of a free skin graft

A different concept was launched by Jossierand (1897) using a canalization technique from the ectopic orifice through to the tip of the glans by a cutting troicart, combined with the

formation of an urethra from a non vascularized, tubed Thiersch flap (fig. 10).

Fig. 10: Neo-urethroplasty: interposition of a tubed free skin graft as neo-urethra after a Bouisson (1861) type of chordectomy.
After Josserand (1897).



2.2.5 Principle of the use of scrotal flaps

There are several methods developed which use scrotal skin in hypospadias repair. It can be used either as part of the urethra during the urethroplasty (Landerer and Modelski 1891, Rosenberger 1891, Bidder 1892, Rochet 1899, Michalowski 1963) or as compensation for the shortness of the dorsal penile skin (fig. 11).

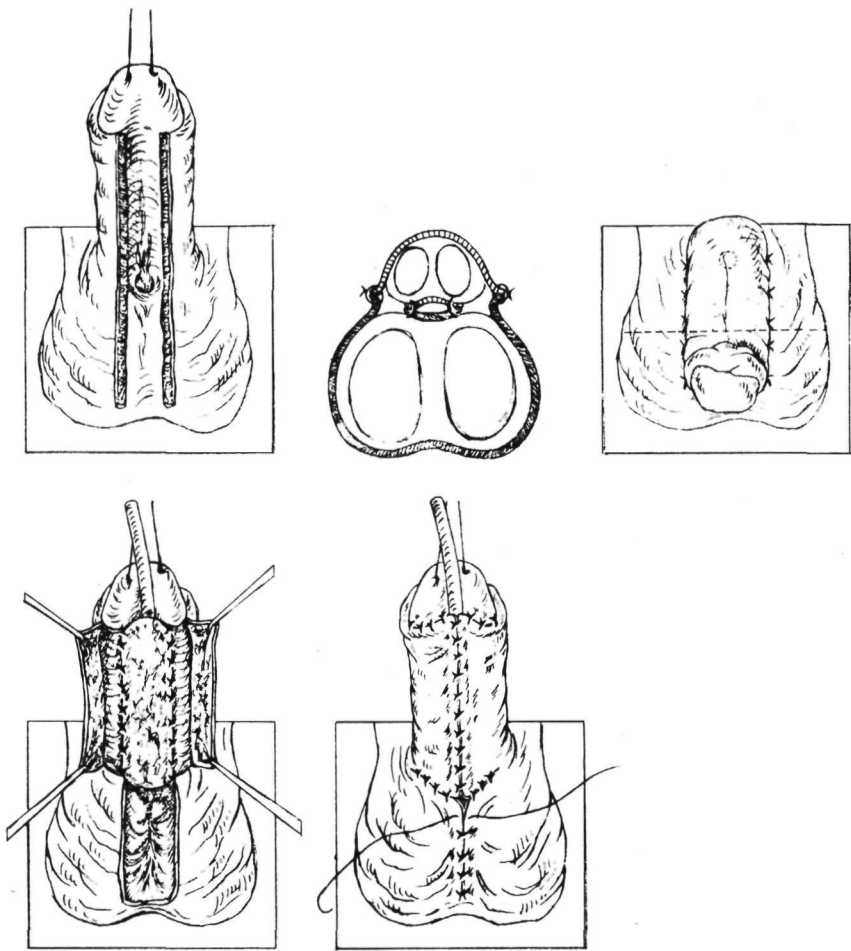


Fig. 11: Neo-urethroplasty: use of scrotally raised flaps for the neo-urethroplasty to build the ventral half of the neo-urethra.

After Rosenberger (1891), Landerer (1891), Bidder (1892).

The latter possibility was initially developed by Beck (1917) and popularized by Cecil (1932), Blair and Byars (1938), Culp (1951) (fig. 12).

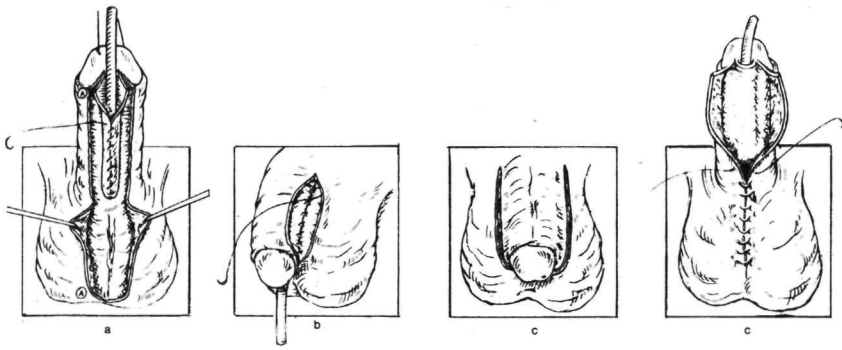


Fig. 12: Hypospadias repair:

the use of scrotal flaps to compensate for the shortness of penile skin at the dorsal side of the penis

- a. neo-urethroplasty (Dyplay I) after previous chordectomy
- b. preparing the scrotal flaps and suture of the penile skin edges to the flaps
- c. third stage: after completed vascularization of the scrotal flap suture, liberation of the penis from the scrotum. After Beck (1917), Cecil (1932).

2.2.6 Principle of urethral advancement

Beck (1900) made use of the elastic properties of the urethra and its intrinsic intensive blood supply in the "urethral advancement technique" for more distally located penile hypospadiac cases (fig. 13). It is still used today (Gross et al. 1970, Waterhouse and Glassberg 1981).

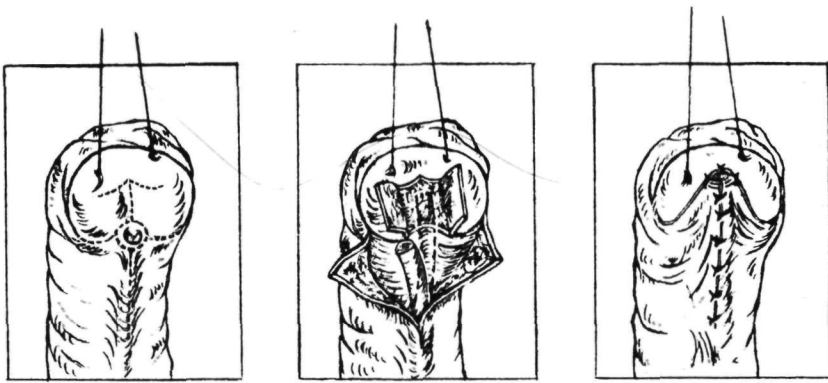


Fig. 13: Hypospadias repair:

principle of urethral advancement by dissecting the urethra

After Beck (1900).

Furthermore, Beck was also the first, who made use of the double layer of the lavish foreskin during the chordectomy by splitting it completely. In this way he created a large skin flap (fig. 14).

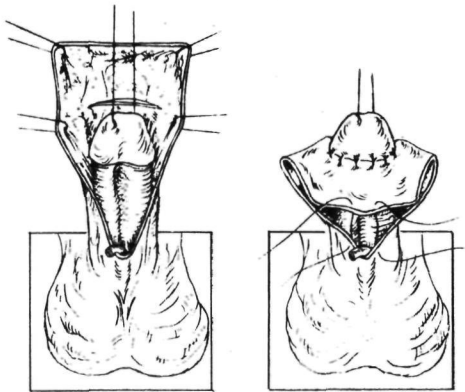


Fig. 14: Hypospadias repair:

splitting of the prepuce in two layers and transposition to the ventral surface through a buttonhole as compensation for the skin shortness.

After Beck (1917).

He used the foreskin as a depot of skin that subsequently was used for the neo-urethra in the second stage. Blair and Byars (1938) use the same principle. Ombredanne (1932) popularized this manouvre in Europe, but used it only as skin coverage after the neo-urethroplasty was performed.

2.2.7 Principle of the tubed, vascularized flap

Mayo (1901) published a method, in which the neo-urethra was formed from a vascularized tubed flap from the prepuce. A similar technique had already been used by Hook (1896). (fig. 15)

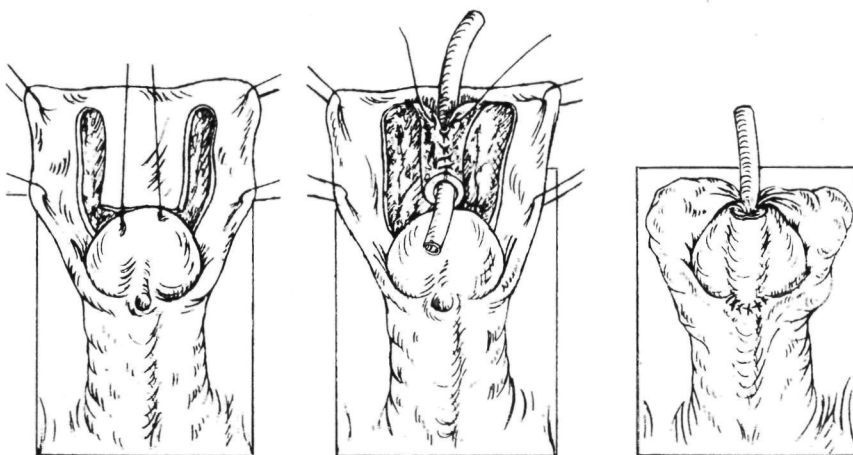


Fig. 15: Neo-urethroplasty:

the use of a vascularized inner preputial flap for urethroplasty. After Hook (1896), Mayo (1901).

It was the first time that the urethra was build up in this way. This made him the predecessor of many surgeons half a century later, who reverted to this technique in developing one-stage repairs. His description of the method was so complicated, that it received remarkably little imitation. In our country van Capellen (1923) reported several successful corrections with this technique.

2.3 Two-stage procedures for hypospadias repair

based on (a combination of) basic methods

Based on the described basic methods a large number of two-stage repair techniques for hypospadias correction have been developed. In the subsequent section we will discuss the most frequently used methods. They are based on either the buried epithelial strip technique (Duplay II) or a fully tubularized neo-urethra (Duplay I).

2.3.1 Methods based on the use of a buried epithelial

strip (Duplay II)

Very popular is the method described by Browne (1936). He performed a type of chordectomy like the one described by Edmunds (1913)(fig. 8). In the first stage the prepuce was swung to the ventral side of the penis to cover the surface after completion of the chordectomy. The second stage is performed at the age of 4 to 6 years. In the description from the early thirties he still uses the Thiersch type of repair with two undermined asymmetrical, sliding flaps like Anger (1874). In the following years he gradually stopped undermining the epithelial strip and only took care to provide a separation as wide as possible between the then buried epithelial strip and the covering skin. Initially a dorsal relaxation incision was used to prevent tension on the sutures (fig. 16).

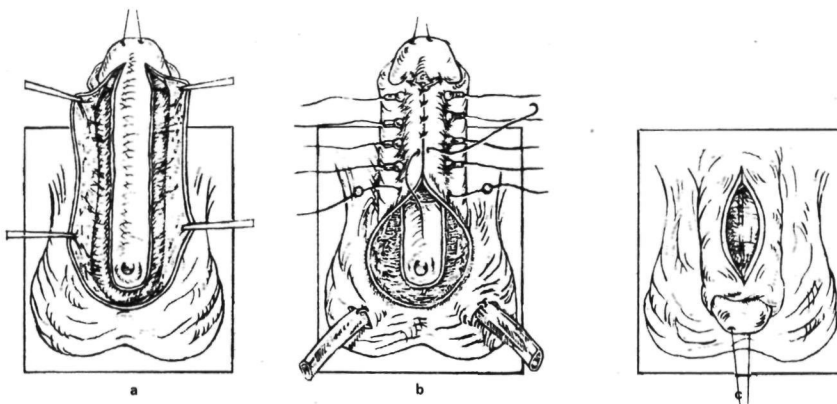


Fig. 16: Hypospadias repair: after the first stage (chordectomy and transposition of the unfolded prepuce)

- a,b. the formation of the neo-urethra by a buried epithelial strip
 - c. dorsal relaxation incision
- After Browne (1936)

This procedure was supported by animal studies which proved, that a buried epithelial strip will form a tube spontaneously (Davis and Traut 1926, Nesbit et al. 1950). Despite the high frequency of fistulas and the formation of a very tortuous and unequally calibered urethra, this method is still widely used. The major progress in reducing the fistula rate was made by the surgeons who recalled the Landerer (1891) procedure and its variations (Cecil 1952, Michalowski et al. 1970). In our country, van der Meulen (1964, 1982) promoted a method in which the volume deficiency at the ventral side was corrected by an Y-V rotation flap technique, thus avoiding disturbing suture lines which may compromise the future urethra. Seldomly a chordectomy was felt to be necessary. The closure after creating the buried epithelial flap was done in at least two layers, using intracutaneous stitches with superimposed skin closure. This gave an important reduction of the fistula rate (fig.17).

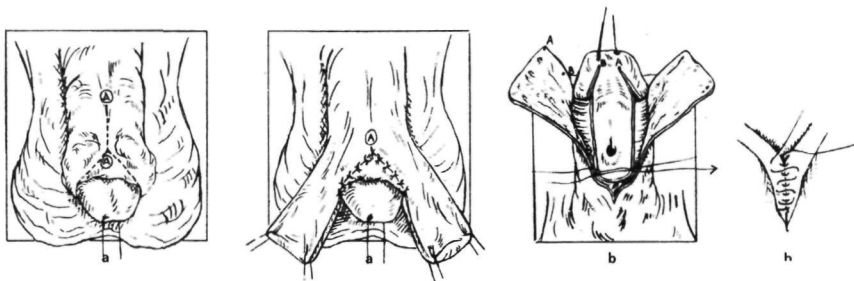


Fig. 17: Hypospadias repair:

- a. transposition of the unfolded prepuce by an Y-V plasty
- b. formation of the neo-urethra by a Browne's type of repair

After van der Meulen (1964).

Farkas (1967) developed another method to reduce the fistula rate. In addition to the buried epithelial strip a split skin graft was used to form a complete tube (fig. 18).

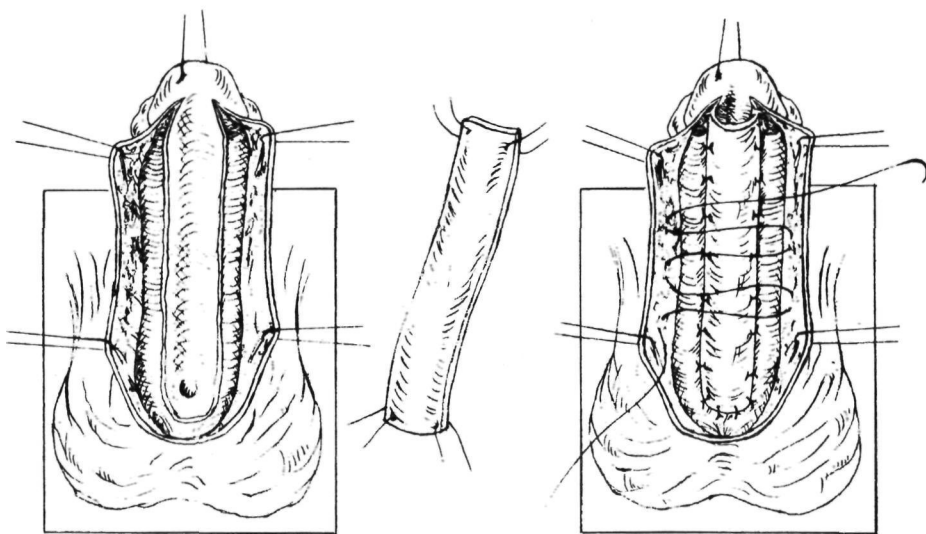


Fig. 18: Neo-urethroplasty: using free skin graft to complete the neo-urethra.

After Farkas (1967).

2.3.2 Method based on the use of a tubed neo-urethra

Smith (1973) experienced to many disadvantages in the original Browne method and reverted to the original Duplay's I method. The neo-urethra is covered in a two layer technique by de-epithelializing the underlying flap (Thiersch technique)(fig. 19).

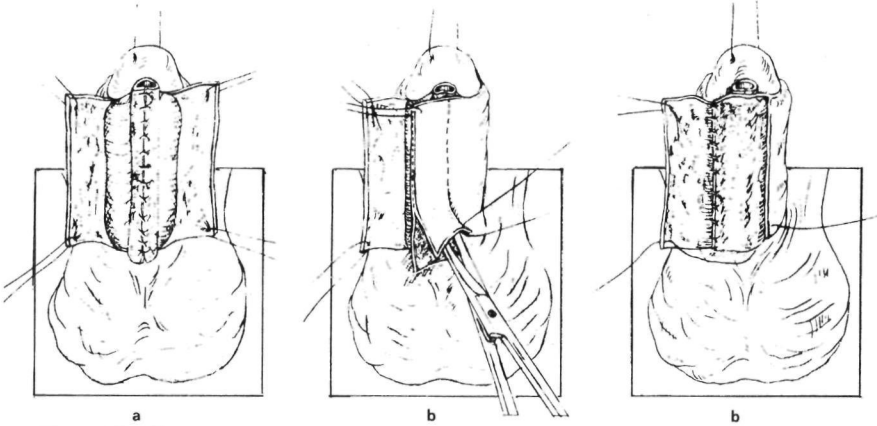


Fig. 19: Hypospadias repair:

- a. neo-urethroplasty using the Duplay I type of repair
 - b,c. covering the neo-urethra using the Thiersch principle of inbricated flaps, de-epithelializing the underlying one
- After Smith (1973).

A balanced, simple and reproducible concept was given by Byars (1951), combining the straight-forward chordectomy procedure of Blair and Byars (1938), with a Duplay I urethroplasty, covered with asymmetrical raised Thiersch flaps (fig. 20).

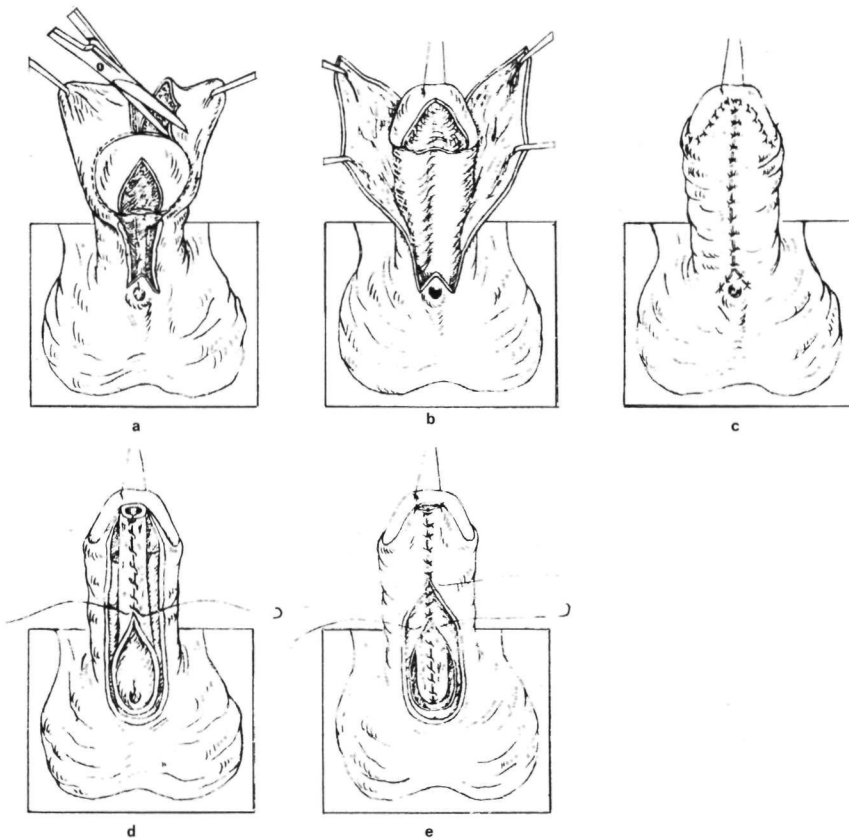


Fig. 20: Hypospadias repair:

a,b,c. Bouisson (1861) type of chordectomy with coverage of the ventral aspects of the penis by asymmetrical raised flaps of the unfolded prepuce (Blair and Byars 1938)

d,e. neo-urethroplasty in a variation of the Duplay I technique (Duplay 1874)

After Byars (1951, 1955).

This technique was a combination of very reliable surgical techniques giving reproducible, acceptable results in many different hands.

These above mentioned methods based on a thorough chordectomy in a first stage and a neo-urethroplasty covered with different raised flaps (preferable in more layers) in a second stage, give a sufficient armamentarium to complete a rather satisfactory hypospadias repair. The method of Jossierand (1897), using free

skin grafts has only survived as part of in a one-stage procedure (see page 56).

Bevans' (1917) method of tunnelization of the neo-urethra combined with the Nesbit (1941) type of chordectomy, was used again by Fuqua (1973) and especially popularized in the USA. Mays (1973) combines this variation of chordectomy in the same session with the forming of the glandular urethra. The rest of the neo-urethra is then subsequently build up in a second stage.

2.4 One stage procedures

Based on the assumption that in most cases of distal hypospadias a chordee was absent, or of minor importance, one stage procedures have been used for the correction of this type of malformation (Beck 1900, Bevan 1917, Matthieu 1932, King 1970, 1981). However, follow-up studies showed remarkably high percentages of persistent chordee, that tended to aggravate after puberty (Sørensen 1953, Farkas 1967, Culp and Roberts 1968). Based on these observations it is obvious that a one-stage repair technique in any hypospadiac case should include a thorough chordectomy.

2.4.1 Using skin

Desprez et al. (1961) described, after a simultaneously performed chordectomy, the use of an elevated skin strip from one of the lateral wings of the prepuce, that was tubed over a No 10 or 12 Foley catheter. The new tube is then rotated in such a way that the suture line is resting at the corpora.

In this way he created the greatest possible distance between the cutis and sutures of the neo-urethra to avoid fistula formation. The new orifice is fixed at the coronal level and the rest of the prepuce is, after being unfolded, wrapped around the penis (fig. 21).

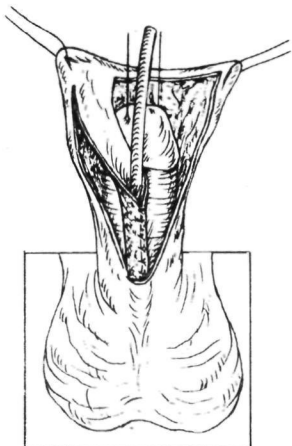


Fig. 21: Hypospadias repair:

One stage technique After Desprez et al. (1961).

Broadbent et al. (1961) created after chordectomy a strip of epithelium originating cranio-laterally of the ectopic orifice. This strip was formed into a tube over a No 12 Fr. catheter and sutured in position through a glandular tunnel or layed into the split glans to cover it with the prepared lateral wings. In doing so they avoided with this technique the proximal anastomosis, which has a tendency to shrink in diameter (fig. 22).

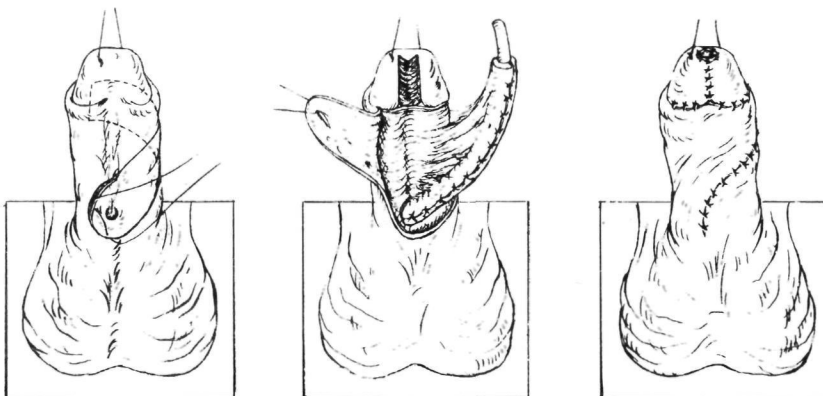


Fig. 22: Hypospadias repair:

One stage technique After Broadbent et al. (1961)

Devine and Horton (1961, 1973) published a different type of one stage repair. After a thorough chordectomy, they created a new urethra from a free graft of the inner layer of the prepuce. In circumcised cases they used skin from the upper inner arm. Both the auto-urethra and the neo-urethra are bevelled and a spacious proximal anastomosis is established. Then the glans is divided into three parts by a V shaped incision. The triangular midline flap is then anastomosed with the tube and the lateral glandular wings are used to complete the closure. The ventral aspect of the penis is then covered with preputial skin flaps in the Om-bredanne (fig. 9) fashion or by the Byars (fig. 20) flap technique (fig. 23).

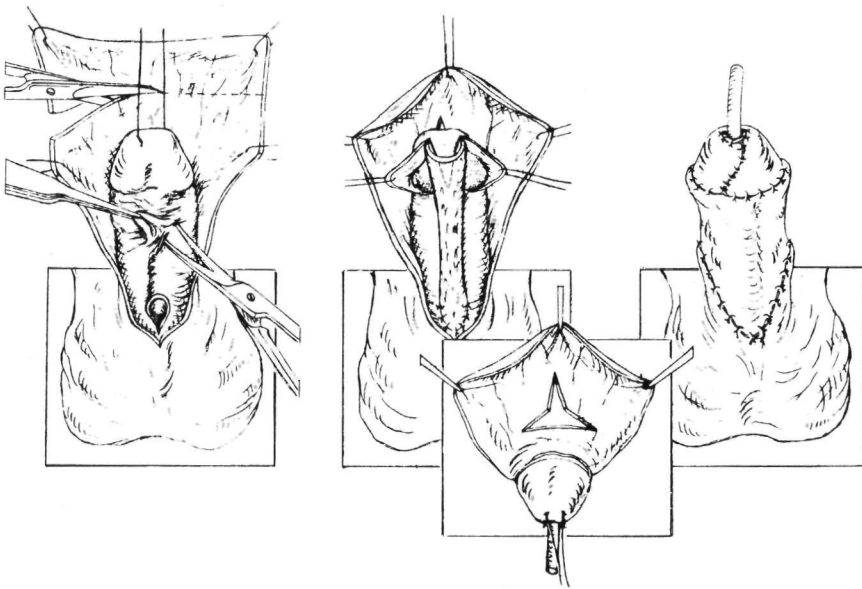


Fig. 23: Hypospadias repair:

one stage technique, combining a tubed free skin graft as neo-urethra with the glandular flip-flap procedure After Devine and Horton (1961, 1973).

This so called glandular flip-flap procedure (1961), has become very popular as a separate procedure for the repair of the distal hypospadias (fig. 24).

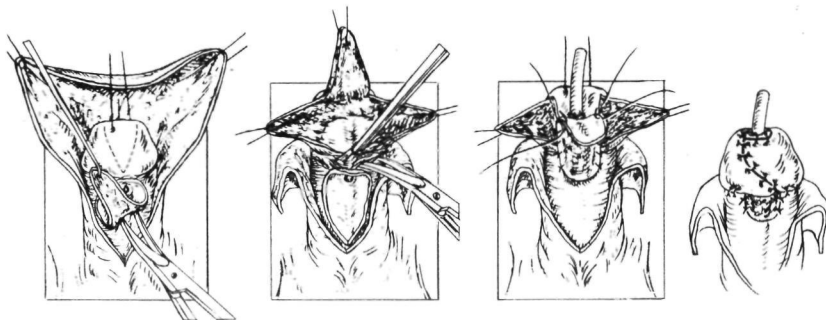


Fig. 24: Hypospadias repair:

one stage technique, the glandular flip-flap procedure
After Devine and Horton (1961).

Mainly because it also allowed minor chordee corrections it is useful for the majority of subglandular distal hypospadias.

In the repair for distal hypospadias without chordee this flip-flap technique can be replaced by the Magpi method (meatal-advancement and glanuloplasty). The operation is easier to perform, and can be done on an outpatient basis without fear of fistula formation (Duckett 1981)(fig. 25).

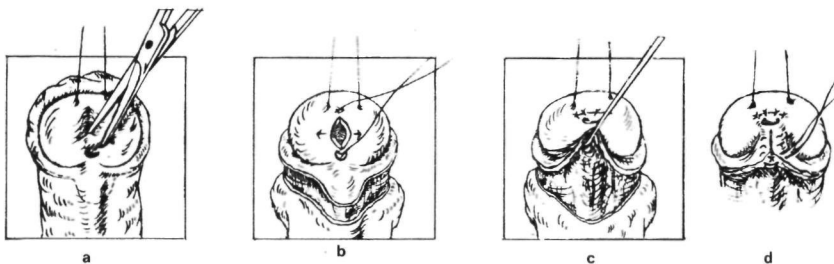


Fig. 25: Hypospadias repair:

one stage technique, the MAGPI procedure

a,b. meatal advancement

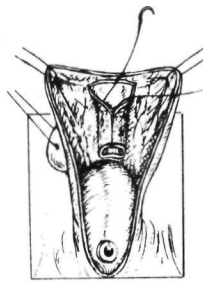
c,d. glanulo-plasty

After Duckett (1981).

Hodgson (1972) described three variations on this one-stage hypospadias repair technique. In the mild cases he used a tubed flap from the inner prepuce layer, in combination with a buttonhole technique at its base to transport the tube from dorsal to ventral. The connection at the glans level was made with a glandular flap (Type I, fig. 26), or the flap was used (according to Matthieu 1932) to form only half the circumference of the distal urethra after dividing of the glans in two lateral parts (Type II, fig. 26). The more proximal hypospadias were repaired with a tubed flap from the outer layer of the prepuce in combination with the buttonhole technique. The orifice was accepted at coronal level, (Type III, fig. 26).

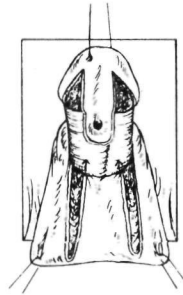
Hodgson type I:

Chordectomy and bridging the short gap of the distal urethra by a tubed island flap of the inner prepuce, delivered to the ventral aspect by a buttonhole



Hodgson type II:

Chordectomy and bridging the gap of the distal urethra, for the ventral half by an island flap of the outer prepuce, delivered to the ventral aspect by a buttonhole.



Hodgson type III:

Chordectomy and bridging the longer gap of the distal urethra by a tubed island flap of the outer prepuce and penile skin, delivered to the ventral aspect by a buttonhole.

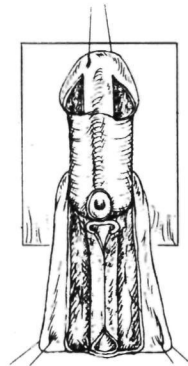


Fig. 26: Hypospadias repair: one stage technique
After Hodgson (1972).

Using the preputial tissue as a tube, Asopa et al. published the first experiences with this method in 1971. This method is a logical consequence of the Desprez method. The flap of the inner

prepuce is raised on its own pedicle but still adherent to the outer layer that served as a cover after anastomosing the tube (fig. 27).

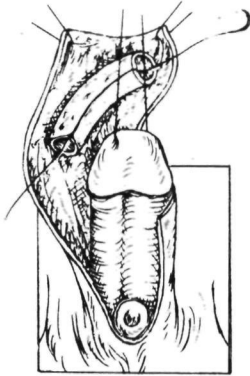


Fig. 27: Hypospadias repair:

one stage technique, using a vascularized flap of the inner prepuce After Asopa et al. (1971).

Standoli (1979, 1982) was the first to raise a flap of the outer prepuce and separate it completely from the inner layer (fig. 28).

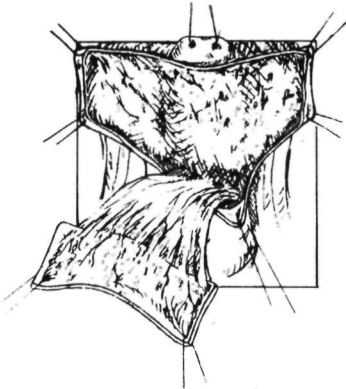


Fig. 28: Hypospadias repair:

one stage technique, using a vascularized island flap of the outer prepuce After Standoli (1979, 1982).

This flap was a real pedicled island flap which was swung around the penis to its ventral aspect. It was then anastomosed with the urethra after chordectomy. The glandular part was placed in the

center of the glans after preparing a good vascular bed by splitting the glans into two parts and excising a sliver of tissue in the center (Barcat 1973). He is also one of the first to perform, during operation, repeated artificial erections, in order to be able to control for the total eradication of the chordee (Gittes and Mac Laughlin 1974). He reported that in some cases, a Nesbit procedure (1965) was necessary to achieve a total correction of the chordee.

Waterhouse and Glassberg (1981) introduced a technique in which the urethra was prepared free from its surrounding tissue and from the subcoronal areas, up to the bulbar part of it. Because of the rich blood supply of the urethra this technique does not lead to necrosis. By stretching the urethra, the gap between the ectopic orifice and the tip of the glans is bridged. This procedure revived an old procedure (Beck 1900). There were other surgeons who followed this procedure with success (Belman 1977, Koff 1981, Ti Seng Chang 1984).

A more recent description of a one stage repair using the prepuce island flap procedure was given by Duckett (1980, 1981). This method, with some modifications, is the same as we used and which is described in detail in Chapter III.

2.4.2 Using tissues other than skin

Li et al. (1981) described a procedure using bladder mucosa to form the neo-urethra. Free grafted tissue has been used before in hypospadias repair. Schmieden (1909) reported a repair using ureter. Tanton (1910) and Adlercreutz (1918) described the successful use of the vena saphena magna as urethral replacement. Axhausen (1918) used the appendix in two cases, and Memmelaar (1947) was the first to report the use of the bladder mucosa as a replacement for the urethra. Rivoir (1954) took a pedicled flap of a tunica vaginalis, using its scrotal skin attachment for skin coverage of the ventral penis. Biebl (1951) preserved the ureter of a patient in which he performed a nephrectomy for other reasons and used it to form a new urethra, leaving the vascular supply intact. Although, recent reports still advocate the use of

such tissues as bladder mucosa (Coleman 1981), we think that these kind of repairs should not be performed anymore.

The transverse inner preputial island flap technique.

3.1 Introduction

One stage hypospadias repair have been performed over a period of time that has been long enough to be able to judge the feasibility of this kind of surgery. The advantages of these procedures will be discussed in detail in chapter V. Since 1980 we use the transverse inner preputial island flap repair in the majority of our hypospadiac cases.

3.2 Preoperative management

The patients are admitted to hospital the day before the operation is scheduled. After a thorough physical examination to evaluate the present situation blood samples are taken for the determination of serum creatinine, blood urea nitrogen, the concentration of hemoglobin and blood cross-matching. Urinalysis completes the investigations. At the day of operation the external genitalia are at least once washed with Povidone-iodine. The prescribed premedication is administered.

3.3 Operative procedure

3.3.1 Anaesthesia

After induction of the anaesthesia by an inhalational anaesthetic mixture or by intravenously administered narcotics, the patients are endotracheally intubated and positioned supine

on the operation table. A loco-regional blockage of the sensible nerves is in most cases applied at the end of the operation.

3.3.2 The standard procedure

The patient is washed again with Povidone-iodine. If the patient is postpubertal, the genitalia are shaven and washed again. After applying the operation sheets the foreskin is freed from the glans by blunt dissection (loosening of the usual adhesions). The glans is then cleaned again. An artificial erection is then created by injecting saline into the corpora with a 23 gauge or 25 gauge butterfly needle (fig. 1). A tourniquet is only necessary in adult patients.

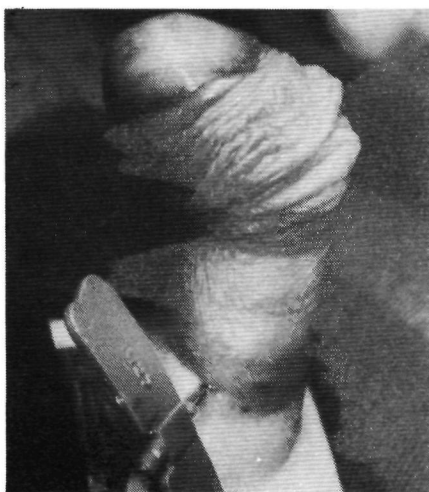


Fig. 1: The artificial erection.

At this stage we decide which kind of operative correction is necessary.

We then place a 3-0 atraumatic Prolene[®] stitch through the glans serving as a holding suture throughout the operation and using it afterwards to fasten the splint. The incision line is marked with a dermatographic pencil encircling the glans and around the meatus extending in the midline proximal until the normal raphe is encountered (fig. 2).

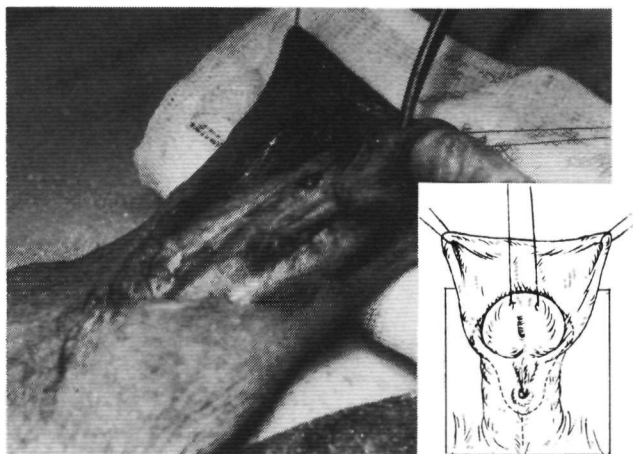


Fig. 2: Circumcision around the corona and the ectopic orifice.

With a knife only the dermis is cut. The subsequent dissection planes are developed using a pair of fine round-tipped scissors. This manoeuvre frees the penile skin easily from the dorsal corpora preparing in a relatively avascular plane of very loose connective tissue and saving the whole nervous and vascular supply of the corpora. We then develop from lateral a dissection plane between the covering thin skin and the distal urethra. Using this lateral approach, it is always possible to dissect this part of skin free from the urethra. At this stage the whole skin is freed from the underlying tissues except for the lateral attachments equidistant to the midline where the remnants of the urethral plate insert into the glandular tissue. These attachments differ from case to case in their distance from the midline. Sometimes two real columns of spongiosa-like tissue can be identified. Before cutting them we electrocoagulate these structures because they are always very well vascularized and can cause profuse serious bleeding.

In the next step the distal urethra is mobilized from the underlying chordee area after insertion of a 10 French catheter (fig. 3).

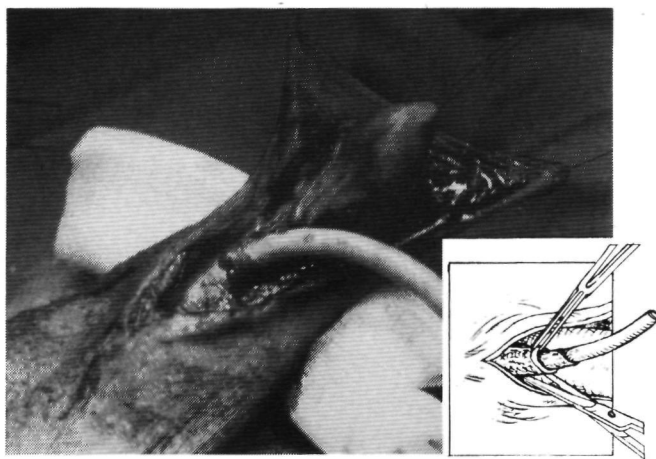


Fig. 3: Starting the urethral mobilization.

The urethra is freed up till normal spongiosal tissue, that surrounds the whole urethra, is encountered. The urethra distal from this point has to be sacrificed because every attempt to make an anastomosis with urethra not surrounded by healthy spongiosal tissue is doomed to fail. In cases where normal spongiosal tissue is found up to the border of the ectopic orifice usually little or no chordee tissue will be present. In these cases another corrective procedure, that only corrects the ectopy of the orifice, may be chosen.

Chordectomy

The following step is the removal of all chordee tissue. This tedious and time consuming process is very important for the final outcome of the whole operative procedure. We start this step at the proximal part of the penis on the point were the spongiosal corpus is completely intact. To prevent unnecessary bloodloss we leave the chordee tissue attached to the last possible moment to the subglandular area where most of the communicating vessels with the corpora are encountered (fig. 4).

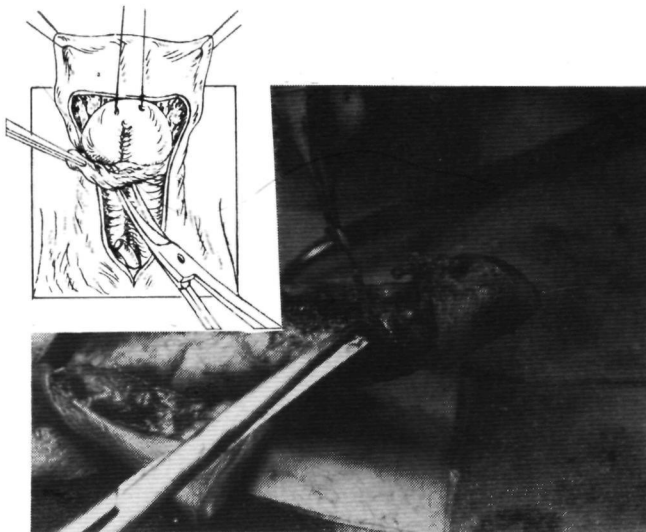


Fig. 4: The chordectomy.

After this procedure a normal urethra should be visible proximal of the original site of the orifice and two glistening blueish corpora separated by a deep groove.

Then a control artificial erection is necessary to see whether there exists still additional bending of the penis (fig. 5).



Fig. 5: Control artificial erection.

If so, additional attempts of removing every bit of constricting tissue are undertaken and the dissection is extended laterally on

both sides. Thereafter again an artificial erection is induced. If still a persistent chordee is seen, a deep longitudinal cut is made between the corpora, this will lead to an outward rotation especially at the level of the most severe bending. This will diminish the curvature for 10 to 15 degrees (fig. 6).

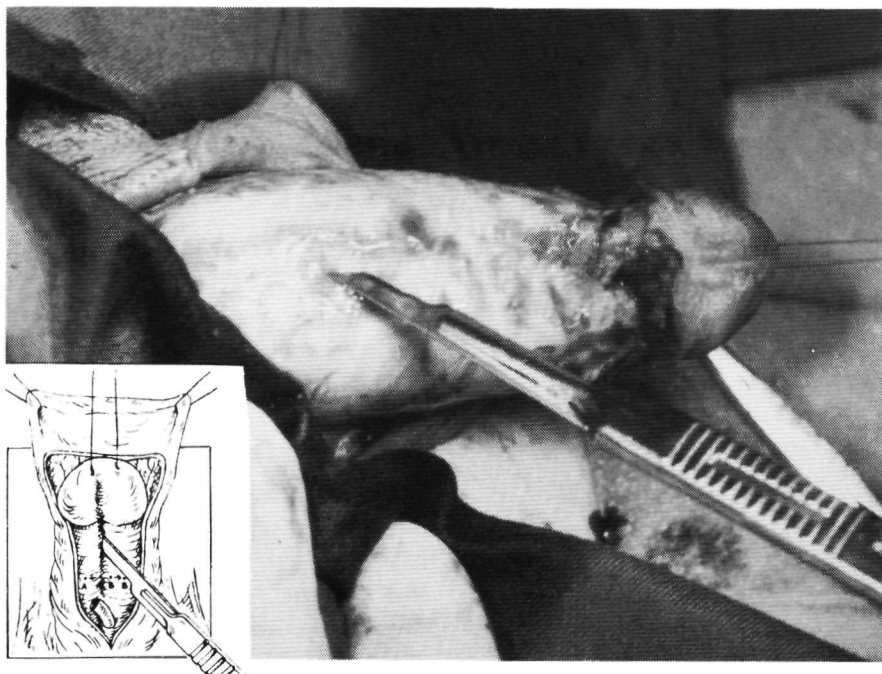


Fig. 6: Longitudinal incision between the corpora

The neo-urethroplasty

After achieving a straight penis the missing piece of urethra is measured and a vascularized neo-urethra is created from the prepuce (fig. 7). The prepuce is put under enough tension by four holding sutures to create a flat surface of the inner prepuce. A rectangular flap of sufficient length is then outlined (fig. 8).

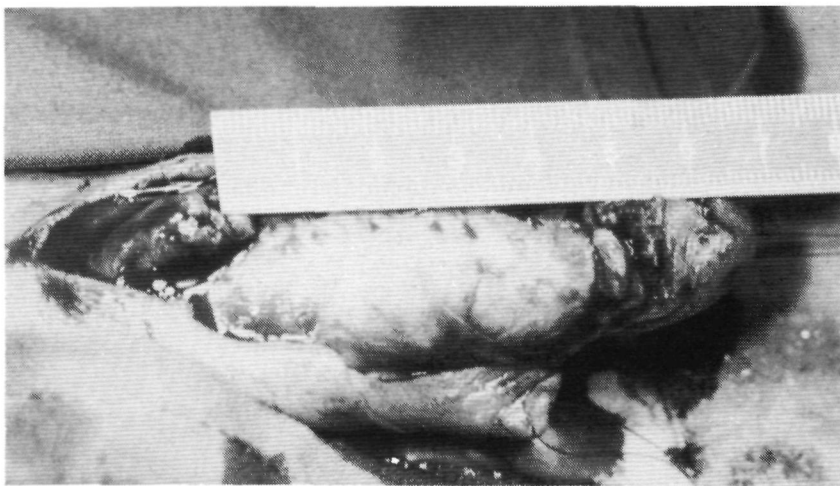


Fig. 7: The missing piece of urethra is measured.

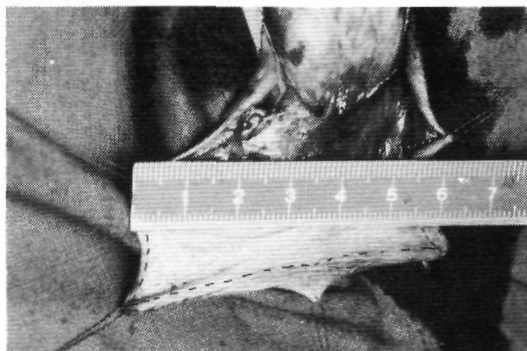


Fig. 8: Measurement the rectangular flap of the inner prepuce.

As diameter for the neo-urethra we take three French more than the calibre of the patient's own urethra. The epidermal layer of the flap is then incised with a knife, meticulously and subsequently its vascular supply is dissected with a pair of roundtipped scissors. In the first instance it is sufficient to dissect a vascular pedicle of about one centimeter. The flap is then tubularized over a catheter with a previously determined caliber (fig. 9).

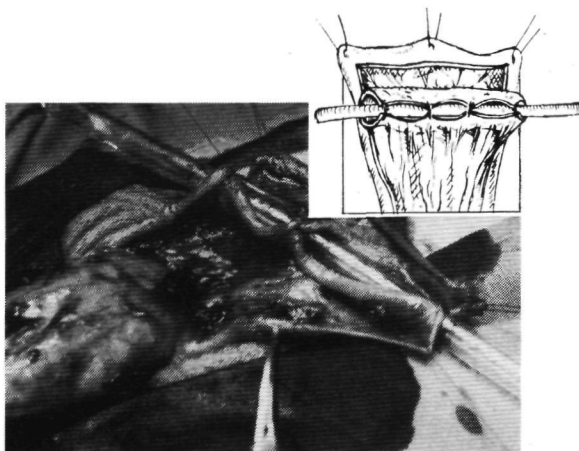


Fig. 9: Tubularization of the vascularized flap of the inner prepuce.

For closing the flap we use an inverting, interrupted suturing technique with 6x0 or 7x0 Polydioxane (PDS[®]) on a small round needle (C2). If the edges of the flap are not closed watertight we use an additional running suture of 7x0 Polydioxane. After completion of the tube the pedicle is freed from the penile skin over a greater distance (fig. 10).

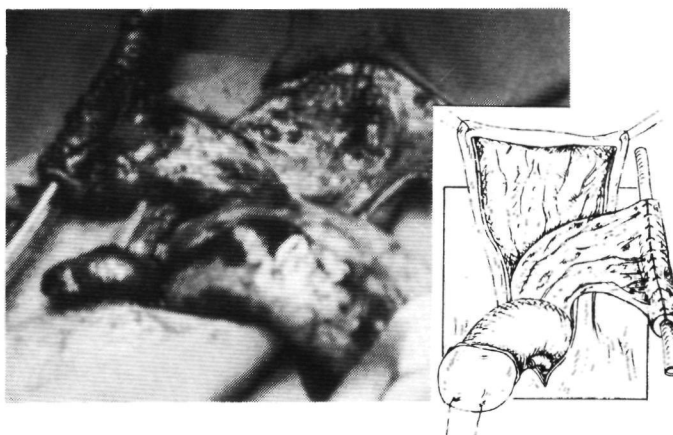


Fig. 10: Freeing up of the vascular pedicle.

During this procedure a check is made, by rotating the tube

between the urethra and the future glans channel, whether this can be done without causing a torsion of the penis. Great care must be taken to preserve the pedicle even at the cost of the penile skin. The latter has proven to possess remarkable regeneration possibilities. The neo-urethra is then transferred from the dorsal to ventral aspect of the penis. It is of utmost importance that the suture line of the urethra will rest on the inter-corporal groove in order to prevent fistula formation. Subsequently a suprapubic bladder catheter is inserted, eventually after filling of the bladder with saline via a 6 French catheter. The Cystofix[®] catheter is fixed with a silk suture to the skin. After this suprapubic bladder drainage the two anastomoses of the neo-urethra are performed. Since a tourniquet is necessary if one starts with the glandular anastomosis we start the proximal anastomosis with the urethra. This is done after we replace the catheter used for creation of the tube by a soft multiperforated silicone stent. This will extend just proximal of the future anastomosis, peripherally of the sphincter. The urethra is than bevelled at its corporal aspect and the neo-urethra at its "antimesenterial" border in order to create a wide oblique anastomosis (fig. 11).

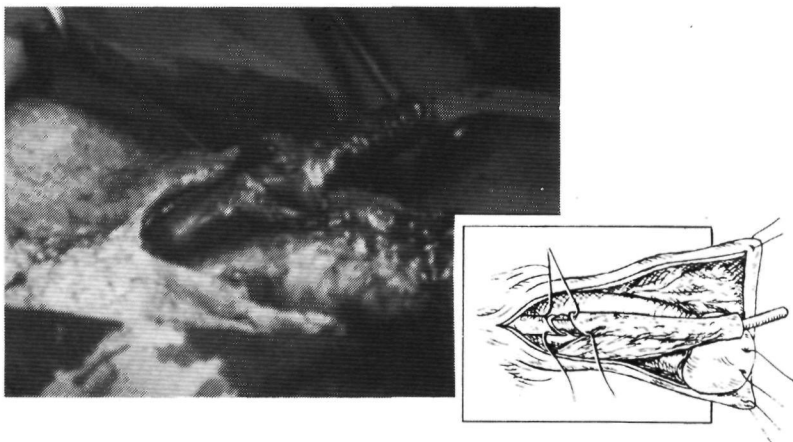


Fig. 11: Creation of a wide oblique anastomosis between the urethra and the vascularized tube.

This anastomosis is made by interrupted 6x0 Polydioxane sutures. The stitch in the corporal corner should include some superficial corporal tissue in order to anchor the anastomosis. The urethra is then fixed to the corpora with widely separated stitches of interrupted 7x0 Polydioxane. At the anastomosis the pedicle is fixed subcutaneously at a more proximal level to prevent fistula formation. Subsequently the proximal anastomosis is established, after the creation of a glans channel.

This channel is created by dissecting in the plane between the corpora and the ventral part of the glans. The proper place for the future meatus is marked and an ellipsoid pastille of the glans epithelium is excised. The long axis of this ellipse should run in the anterior-position in order to create a more physiological appearance. With this manoeuvre it is possible to perforate the glans completely with a pair of scissors from below (fig. 12).

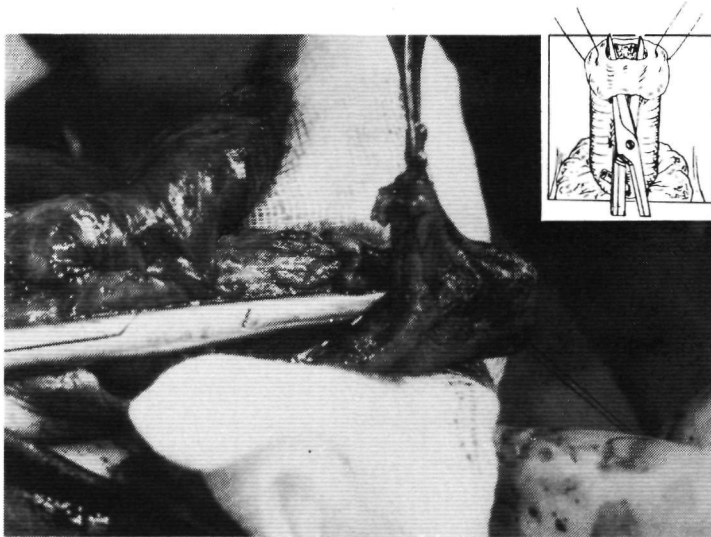


Fig. 12: Creating of the glans channel.

It is always necessary to remove quite an amount of glandular tissue, especially from the ventral side. This should be sufficient enough to allow the free passage of an 20-22 French Beniqué even in children (fig. 13).

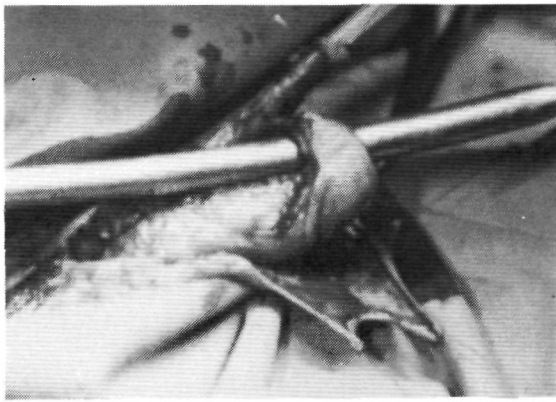


Fig. 13: Measurement of the width of the glans channel.

After the urethra is passed through this channel, it will be trimmed to the appropriate length. The anastomosis is made with four to six interrupted 6x0 Polydioxane sutures.

Penile skin preparation to cover the ventral surface

The dorsal penile skin is then stretched out and under guidance of illumination a vertical incision is made in a plane which is as avascular as possible. This incision usually needs to extend just proximal of the dogears site. The dorsal aspect of the remnants of the innerlayer are then anastomosed with the incised penile skin (fig. 14).



Fig. 14: Creation of the penile skin flaps.

The flaps of the prepuce are brought around to the ventral aspect and closed in the midline or in a Z plasty fashion. The skin is approximated with interrupted 5x0 Vicryl[®] sutures. No drainage is used (fig. 15).

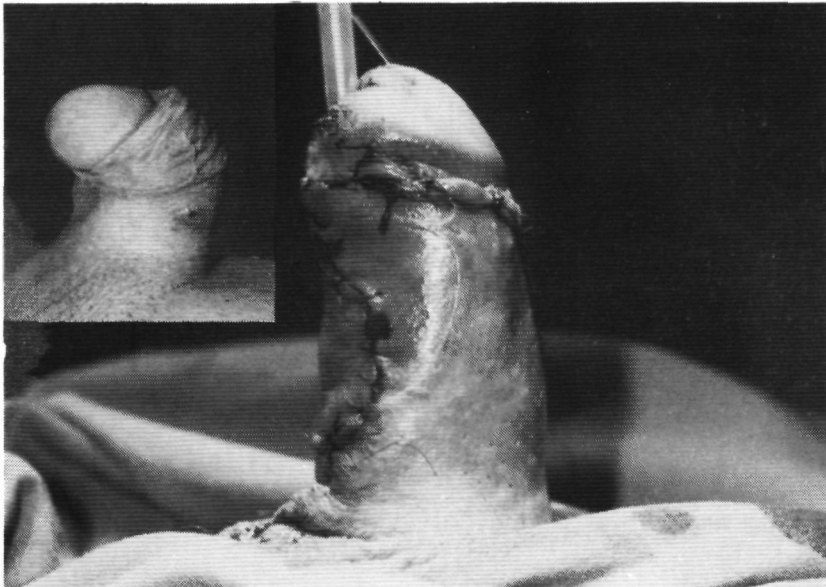


Fig. 15: At the end of the operation.

The inset shows the preoperative appearance.

The operation is then finished by cleaning the operation field with a chlorhexidine solution and application of the dressing. The dressing is not only to be considered as the end of the operation but also as the beginning of an uneventful recovery!

The type of dressing we use is the Silastic Foam[®] dressing consisting of two components which, when mixed together, cause a polymerisation of the elastomer. The catalyst (0.6 ml/10 ml elastomer) is added to the elastomer and the material is mixed thoroughly for 15 sec. Then the mixture is poured around the base of the penis within a tube-like mould. Any piece of smooth plastic can be used for this purpose. The mixture then starts to polymerise and expands about four times its initial volume. The result is a soft pliable dressing that immobilizes the penis (fig. 16).

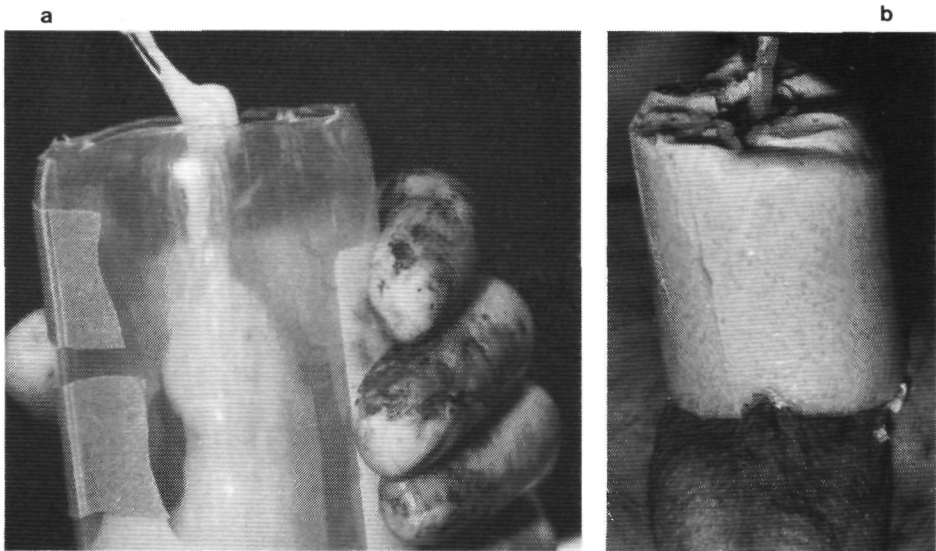


Fig. 16: a. The mixture is poured around the penis
within the plastic mould
b. A supple soft dressing

3.3.3 Supplementary procedures

Chordectomy

In some cases there will still be a bending of the penis after scrutinous dissection of all the chordee tissue. This bending is caused by a relative hypoplasia of the ventral part of the cavernosal corpora.

This deformity can only be corrected by an augmentation at the ventral part of the corpora or by shortening the dorsal part of the corpora. It depends on the size of the penis which method we will use. In patients with a relatively small sized penis, we perform a augmentation at the dorsal side of the corpora.

By making a deep transversal cut in both corpora at the level of the most pronounced bending, the cavernosal tissue will be exposed and the defect in the tunica albuginea will become more or less elliptical by stretching the penis till it is straight. This corporal defect will then be sealed off with a free patch of full skin (Devine and Horton, 1977)(fig. 17).

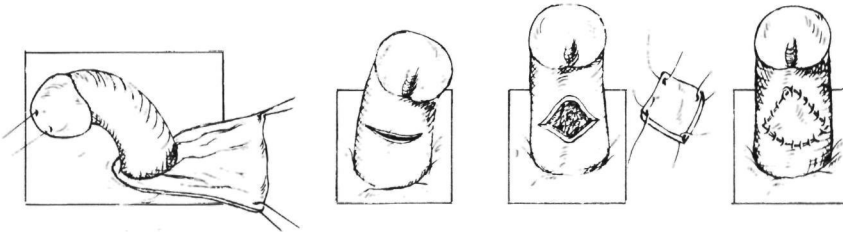


Fig. 17: An interposition with a free flap to correct persistent chordee.

If there is sufficient penile length the Nesbit (1965) procedure is preferable because it is less traumatic. At the dorsal side the maximal bending point is identified and marked. Over a

distance of about 2.0 cm the neuro-vascular bundle is dissected free and kept away laterally (fig. 18).

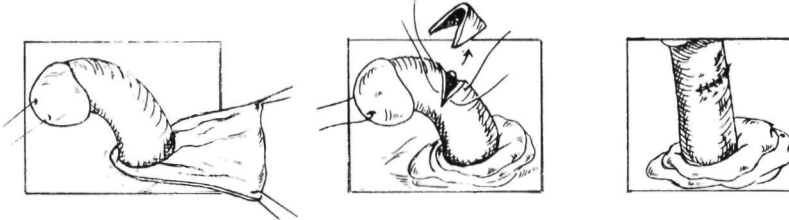


Fig. 18: Nesbit procedure to correct persistent chordee.

The triangular piece can be taken out of inverted, preparing the vascular bundle aside.

Then in each corpus a small cut of 0.6 till 1.5 cm is made and with mattress sutures an invagination of enough corporal tissue can be achieved to correct the bending completely. So we do not remove a piece of tissue as Standoli (1982) reported in his series. As suture material we use a longlasting material of sufficient strength such as 3x0 Polydioxane.

Neo-urethroplasty

In some cases the vascular pedicle of the neo-urethra is too massive to transfer it on one side only. In these cases we will split the pedicle over a short distance in longitudinal direction. Through this buttonhole the penis is then delivered to the dorsal aspect of the pedicle. Positioning of the urethra will then provide half of the pedicle at either side of the penis. This procedure will create a more symmetrical appearance in these cases (fig. 19).

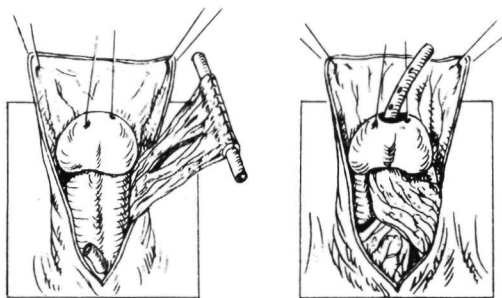


Fig. 19: Dividing the vascular pedicle of the neo-urethra.

3.4 Postoperative management

After the foam dressing is applied, the anaesthesiologist will apply in most patients a loco-regional nerve block. The patients are placed in a lateral knee-chest position. Because the general anaesthetic effect is still working the patient will be motionless.

After preparing the skin over the sacrococcygeal region with an alcohol- iodine solution the anaesthesiologist will puncture the sacral hiatus with a 23 Gauge needle. After checking the proper position of the needle in the caudal canal with a saline injection (absence of subcutaneous swelling, no aspiration of blood or spinal fluid), a dose of marcaine solution will be given into the caudal canal.

Postoperatively the children are allowed to move freely in their beds. Post pubertal patients are for 24 hours confined to their beds. The next day mobilization is started. If the recovery is uneventful the dressing is removed on the third postoperative day. On the seventh to eighth postoperative day the subrapubic catheter is clamped and the patients are allowed to micturate through the penis with the silicone stent still in place. If no

complications occur the next day the holding suture through the glans is cut and a urine specimen is collected. The next day the splint is always spontaneously removed during micturation. Then the suprapubic catheter is removed and 24 hours later the patients are dismissed. If necessary, based on urine cultures, an antibiotic agent is prescribed.

3.5 Discussion

In contrast to various other corrective procedures for hypospadias the inner preputial island flap technique makes it possible to correct the majority of hypospadiac patients (see also chapter V).

Anaesthesia

In our opinion loco-regional blockage of the sensible nerves in the immediate post-operative period has major advantages.

The post-operative pain relief is very satisfactory, and it last for 4 to 6 hours. We did not see adverse effects which have described as complications of this procedure (Schulte-Steinberg 1980). The main advantage, is that the patients, are pain free for a prolonged period of time without being sleepy as with normal analgesic agents. Especially in children it is important that the contact with the parents can be re-established almost immediately after the recovery-room period.

Since the control over the loss of blood can be very strict by using a tourniquet and/or vasoconstrictive agents (Duckett 1981, de Sy et al. 1981) there is no need for controlled hypotensive anaesthesia.

Chordee resection and correction of the penile curvature

Since the extent of the chordee can hardly ever be evaluated preoperatively we think that for this purpose an artificial erection is necessary after the entire penile skin has been released.

Only in some cases a preoperative picture gives sufficient information to judge the severity of the chordee.

The absence of normal spongiosal tissue around the urethra is the best indication for the extend of chordee tissue. In no case we were able to correct the bending of the penis by only removing fibrotic tissue between the ectopic orifice and the glans, if there was no normal spongiosal tissue surrounding the most proximal part of the urethra. Therefore we always dissect the urethra over a distance of at least one centimeter proximally of the point where the spongiosal corpora are completely surrounding the urethra. The intrinsic factors, like the urethralplate and the non-fused mesenchyme on each side that cause the chordee, are ample discussed in chapter I.

Furthermore, although infrequently the bending of the penis can be caused by a hypoplasia of the ventral side of the cavernosal corpora. In this case the bending can only be corrected by an augmentation at the ventral side or by removing a piece of tissue at the dorsal side of the penis.

Neo-urethroplasty

The most ideal replacement for the urethra has not been found yet. Besides good elastic properties it has to be able to respond to androgen stimulation and it should be surrounded by tissue that would prevent a collapse of its lumen during erection. Of all tissues used so far in experimental or clinical situations, the prepuce meets all these demands in the best possible way. It possesses androgen receptors and is very tender with good elastic properties especially on its inner surface, where the stratum corneum is very thin.

However, there is still a dispute about the amount of androgen receptors in human foreskin. In hypospadiac patients there is ample proof that there is at least a sufficient number available to allow a more or less physiological growth of this tissue at puberty (Svensson 1979, Coulam et al. 1983). In culture, fibroblasts derived from human genital skin of hypospadiac patients are able to metabolize testosterone to a greater extent and to bind

dihydrotestosterone specifically to a greater extent than fibroblasts derived from non-genital skin. Based on this observation it is very likely that preputial skin will respond to androgenic stimulation (Kaufmann et al 1978). Moreover, since it has been found that the specific activity of the dihydrotestosterone in the prepuce is concentrated in the germinative layers (Weaker and Sheridan 1983), which part of the prepuce will remain intact during the described operative procedure.

Sometimes, in the operative correction of the so called "hypospadiac cripples", we take advantage of the presence of androgen receptors in the prepuce. If the tissues are of a poor quality, we apply testosterone locally in an indifferent unguent, which will improve the quality of the tissues impressively. Thanks to the augmented bloodflow and thickening of the individual dermal layers it is more likely that the skin will survive yet another surgical procedure.

The advantages of the particular bloodsupply of the prepuce and the surgical consequences thereof are described in chapter IV.

The urinary diversion

There are several methods to divert the urine after the hypospadias repair. There are only few authors who allowed an immediate post-operative voiding through the neo-urethra (Ombredanne 1911, Crawford 1961). They had to rely on the cooperation of their patients not to press too hard at micturation. This makes the method unsuitable for children. Temporary diversion of the urine after operation of the urethra has been advocated by most authors. The main reason for not allowing urine to pass through the operated area is the fear for fistula formation. The suture lines of the neo-urethra at its anastomosal sites will become edematous within a few hours after the operation and not watertight anymore. If urine would pass forcefully some of it will leak at the sutures to the surrounding tissue causing small urinomas, which are not only responsible for possible fistula formation but can also become foci for infection.

The most widely used urinary diversions after the formation of the neo-urethra are:

- a. Transurethral catheter drainage
- b. Perineal urethrostomy
- c. Supra-pubic catheter drainage

- a. Transurethral catheter drainage

The advantages of this method are:

1. easy to perform
2. no extra surgical intervention is necessary

The main disadvantages are:

1. It irritates the trigone of the bladder causing bladder spasms
2. There is a high risk for infections (Shohet et al. 1983)
3. It can cause erosion at the actual repair site
4. It can cause strictures at the penoscrotal corner
5. It sometimes can be difficult to remove the catheter because of the greater circumference of the catheter at the side of the balloon.

This method is used almost exclusively in distal hypospadias repairs, where the catheter stays in only for a few days (Allen and Spence 1968, Novak et al. 1980, Bochiotti et al. 1982, Gonzales et al. 1983). For more complicated repairs only Desprez et al. 1961, Culp and Roberts 1968, Perovic 1983 rely on trans-urethral catheter drainage.

- b. Perineal urethrostomy

The main advantages of this method are:

- 1 A catheter may be left in place for a prolonged period of time, not creating erosion problems.

- 2 The patient can void freely. Especially in the presence of bladderspasms, the urine may pass beside the indwelling catheter and will dribble down the perineum, and does not harm the anastomosis.

The main disadvantages of this method are:

- 1 Especially in children there will be a cystitis within 24 hours due to fecal soiling.
- 2 It asks for a supplementary surgical procedure that bears its own risk of fistula or stricture formation (Vulliet 1905, Marberger and Bandhauer 1965). Marberger found these complications serious enough to forbid the use of this method at his clinic.

This method is widely used by the followers of the Duplay types of hypospadias repair (1874), (Josserand, 1897, Browne, 1936). It has still many advocates in our days (Devine and Horton 1961, Hinderer, 1971, Johanson and Avellàn, 1980, Hendren and Crooks, 1980, Hensle and Goldstein, 1981).

c. Suprapubic catheter drainage

This can be accomplished by using a percutaneous puncturing technique (Rees et al. 1981, Standoli 1982, Belman and Kass 1982, Hanna 1983), or an open operative procedure for inserting a Pezzer or Malecot catheter (Duckett 1980, de Sy and Oosterlinck 1981, Belman 1985).

The advantages of this method are:

- 1 The skin area in which the drainage is established is far away from any possible source of infection.

- 2 The trigone of the bladder is not irritated. So less bladder-spasms will occur. Especially the open procedures allow for an exact positioning of the catheter at the bladder dome.
- 3 The lumen of the drainage canal is much wider than the same calibered Foley catheters.
- 4 It can be removed without possible damage to the repair.
- 5 In case of the percutaneous techniques it can be used at any stage of the operative procedure without any spillage of urine.

The disadvantages of this method are:

- 1 Another surgical procedure is needed.
- 2 Especially with the troicart technique bladder spasms can be encountered more frequently.

We prefer the Cystofix[®] catheter mainly because the very sharp and very thin needle troicart based on the peel off principle (fig 20) causes a minimal surgical trauma.

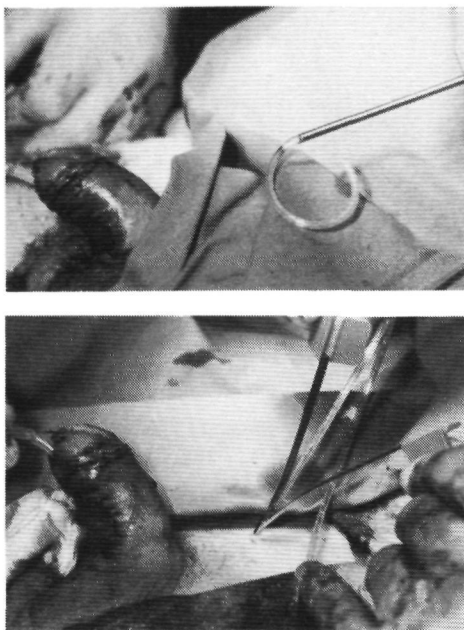


Fig. 20: Suprapubic cystostomy using the cystofix[®] with its memory pigtail and its peel off principle

The poly-urethane catheter with memory pigtail end is inserted into the sharp needle upto the first blue marking point. The skin may or may not be incised. Then the needle is put in, easily passing the skin and fascial barriers. After positioning the needle and its content (a 10 or 15 French catheter) into the bladder the puncture needle can be removed by tearing it into two pieces. The spillage of urine is avoided by connecting the collecting bag at the end of the catheter beforehand.

Drainage through the original ectopic orifice is only of historical interest. Duplay (1874), Jossierand (1897), Charnock (1943), May's (1951), Byars (1951), all used in the first years of their surgical work a separately reconstructed neo-urethra that was stented and let the urinary drainage take place through the ectopic orifice. Most of them turned later to an immediate anastomosis between the neo-urethra and the proximal urethra using one of the aforementioned methods of urinary diversion.

Suture material

Since Lister (1827-1912) had found a method for sterilizing suture material the sheepgut varieties became more widely used. By using tanning procedures the resorption times were prolonged and this chromic-gut sutures became very popular. It took up to the early sixties before new resorbable materials were developed and in 1969 the glycopolyglucolic acids became available.

The most widely used materials are still catgut, (both plain and chromic tanned), the polymer agents like glycopolyglucolic acid and the non-resorbable materials such as polypropylene (Prolene[®])

One of the major disadvantages of the catgut varieties is that the absorption by the body takes place by phagocytosis and denaturation by collagenase. Therefore the strength diminution is hardly predictable. As capillary thread (plain catgut) its use should be avoided in cases where there is a high infection risk (Blomstedt and Osterberg 1978). The same is more or less the case in the polyglactine 910 (Vicryl[®]) and polyglycol acid (Dexon[®]) sutures. The least damage to the tissue is caused by the very fine metal wires or the monofilament polypropylene. The main disadvantages however of these products are that they have to be removed. In the younger age group this would mean another period of heavy sedation or general anaesthesia.

Therefore we prefer to use the monofilament resorbable polydioxane suture PDS[®]. It has good handling properties, secure knotting possibilities and no tendency to fray (Berry et al. 1981). Its resorption time by hydrolysis is substantial and lasts for 30 days upto 4 months (Albers et al. 1982). Its reliability even in case of infection is very high (Alexander et al. 1967). Despite its prolonged exposure to urine, until now we have not observed stone formation in our reconstructive operations using this material.

The use of histo-acrylates uniting the wound edges by a coagulating process never did find a widely spread use (Vahlensieck 1973).

The proper dressing of the wound is not just the end of the operation but also the beginning of an uneventful recovery. Because in the early days of hypospadias repair most patients were post pubertal, much emphasis was given to prevent an erection in the immediate post-operative period. To achieve this many methods have been advocated, using totally concealing or partially concealing dressings, to compress the tissues to prevent edema and hemorrhage and to immobilize the penis. This is for example accomplished by elastoplast over gauzes containing antibiotics, (Culp and Roberts 1968, Hensle and Mollit 1981), or by the use of gauzes soaked in friars balsam (Yarbrough and Johnston 1977). Devine proposes an intermittent irrigation with cold saline of the penis, just wrapped in cotton, every half hour during the first day with changing of the dressing 3 or 4 times (Cromie and Bellinger 1981). In their survey these latter authors reveal that 95 % of the surgeons felt that the dressing had any bearing on the outcome of hypospadias repair. They all described the ideal dressing as compressive without causing ischaemia and necrosis. Most importantly, it should immobilize the penis. By some authors this is even extended to the whole patient, restraining all four extremities for a week (Hendren and Crooks 1980). Most surgeons recommend a complete bedrest for 5 days, although this advice is questionable in young children, if indeed not all four extremities are fixed.

This situation was severely criticized by parents of three children in our survey. Despite newer dressing methods were recommended (Falkowski and Firlit 1980, Vordermark 1982), no method was able to dose the tightness of the dressing in such a way that edema and haemorrhage was prevented without causing necrosis.

To evaluate the pressure characteristics of our dressing with Silastic Foam[®] we performed pressure measurements in our patients.

The pressure measurements were done during application of the dressing, the subsequent polymerisation phase, and during the

first post-operative day.

A microtip-transducer for the pressure measurements was placed between the penile skin and the foam dressing. The pressure measurements over a prolonged period of time were via wireless transmission send to a recording device (fig. 21).

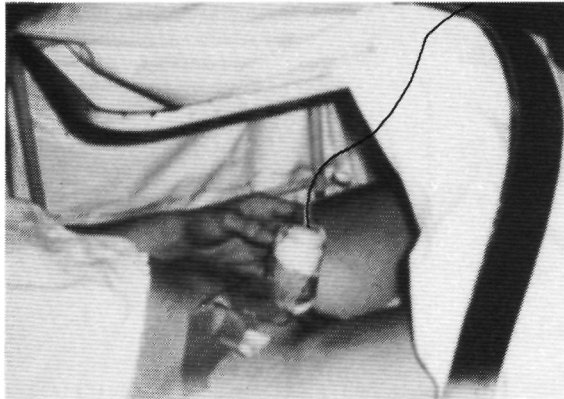


Fig. 21: Pressure measurement using the microtip transducer and a wireless transmission system.

Measurements were performed in four patients and the highest pressure recorded in these patients during the polymerisation phase and of the silastic elastomer was between 42 and 46 cm. H₂O.

The maintenance pressure varied between 24 and 27 cm. H₂O. The same measurements were performed during the application of a "normal" gauze dressing in one patient. Even the gentlest handling of this type of compressive dressing showed pressure rises of over 150 cm. of H₂O with varying maintainance pressures as high as 60-90 cm. H₂O on different spots of the penile circumference.

Therefore, this foam material is in our opinion the ideal dressing after penile surgery, being not adhaerent, circumferential equally compressive with pressure levels just above the venous pressure, not interfering with the bloodsupply, immobilizing the

penis, allowing the patient absolute freedom without the necessity of further immobilization of the patient. This allows mobilization of the children the day following surgery. The removal of the dressing is absolutely painless. We have used it in all our repairs since 1980 with great satisfaction. We learned about this type of dressing from Mattelaer (1980). De Sy and Oosterlinck (1982) reported its use in hypospadias repairs.

Studies on the vascularization of the human penis.

4.1 Introduction

Although there exists a longstanding experience in reconstructive surgery with vascularized flaps, the use of island flaps with long vascular pedicles has been introduced only recently. That vascular pedicled island flaps dissected from the prepuce can be used, is mainly based on clinical experience (Hodgson 1981). The vascularization of the prepuce and its relationship with the penile bloodsupply has been studied mainly in experimental animals. We therefore, performed an experimental study on the vascular architecture of the prepuce and penile skin in humans, with special emphasis on the described method of preparation of the vascular pedicle of the transverse inner preputial island flap (see Chapter III).

4.2 Patients and methods

The vascular architecture of the prepuce and the penile skin was studied at autopsy in six male patients who died of causes not related to the urogenital tract. Four patients died as a consequence of cardio-pulmonary failure, one patient died after a road-traffic accident, and one patient due to massive metastasis of a bronchial carcinoma. The mean age of the patients was 45 years (range: 20-60 years).

At autopsy the vasculature underneath the level of the aortal bifurcation remained intact in all six cases. Also the urinary tract was left intact below the level of the third lumbar vertebra in all but one case. In the latter one the bladder was removed for histological examination since there was macroscopic

evidence for a tumor. In two cases, including the case in which the bladder was removed, the feasibility of the radiological imaging technique was tested (fig. 1).

- a = dorsal penile artery
- b = deep (dorsal) penile artery
with glandular branches
- c = Bulbo urethral artery
- d = recurrent arteries:
connection between external
and internal iliac system

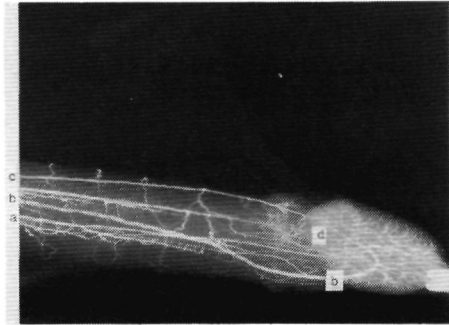


Fig. 1: The penile bloodsupply by the internal pudendal artery

A highly dense radiopaque contrast medium (Barium-sulfate solution) was injected in the common iliac artery on both sides. With this method arteries upto a diameter of 0,3 mm could be visualized, including terminal branches from the internal and external pudendal artery. Subsequent dissection of the penile and preputial skin showed the contrast medium in even the smallest vessels (fig. 2).

a = dorsal penile artery
 d = recurrent arteries:
 communication between
 internal and external
 iliac system
 A = external pudendal
 artery (deeper layer)

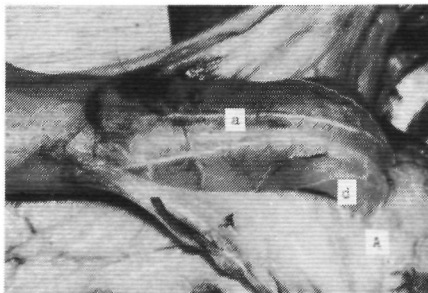


Fig. 2: The bloodsupply of both the penile body and
 the penile skin

In the remaining four cases the contrast medium was administered after the preparation of a transverse inner preputial island flap as described (see Chapter III). The contrast medium was injected in the common iliac artery at both sides and an X-ray study was performed of both the remaining penile skin and the transverse inner preputial island flap.

4.3 Results

In three out of four cases we were able to show contrast medium in the transverse inner preputial flap in a substantial number of small vessels and in some larger vessels in the vascular pedicle (fig. 3).

C = end arteries of the
deeper external
pudendal system
d = recurrent artery
system

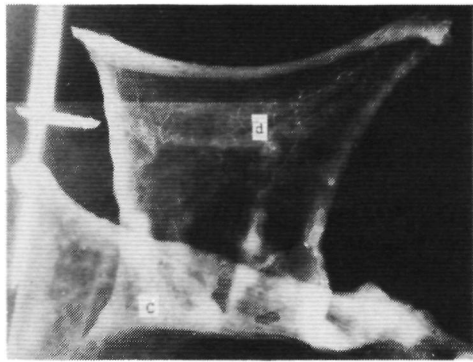


Fig. 3: Bloodsupply of the island inner preputial flap
and its pedicle

Further more it was obvious that the preparation of the pedicled flap did not interfere with the vascularization of the remaining penile skin (fig. 4).

B = superficial layer
of the external
pudendal system
A = deeper external
system (root of
the pedicle)

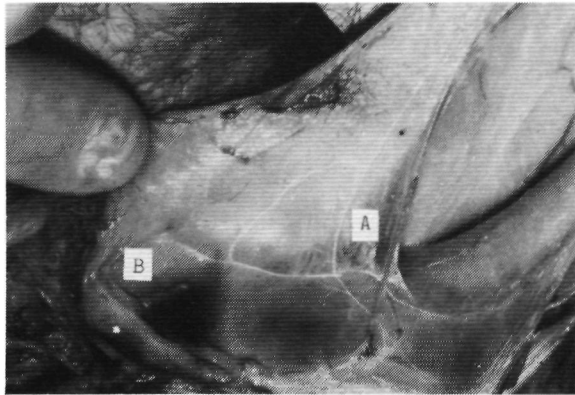
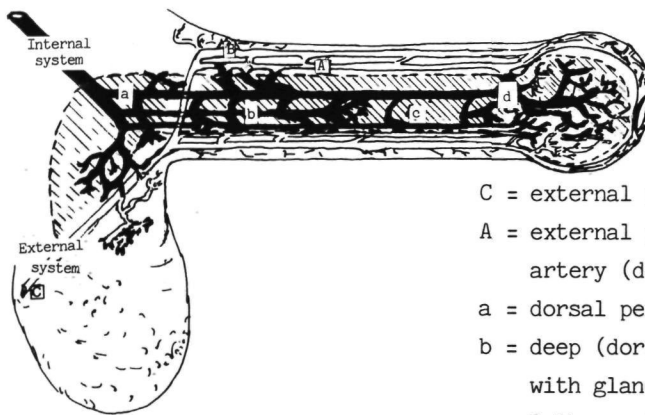


Fig. 4: Bloodsupply of the penile skin

The results of these radiological investigations are schematically summarized in fig. 5, showing the vascular architecture of the human penis.



- C = external pudendal artery
 A = external pudendal artery (deeper layer)
 a = dorsal penile artery
 b = deep (dorsal) penile artery with glandular branches
 c = Bulbo urethral artery
 d = recurrent arteries: connection between internal and external iliac system

Fig. 5: Schematic representation of the bloodsupply of the human penis

4.4 Discussion

The replacement of the missing piece of urethra in hypospadias formed for a long time the main problem in the correction of this anomaly. An eventual repair before the onset of puberty should be performed with tissue that should be able to grow under the influence of androgen stimulation. Furthermore this tissue should also be able to stretch quite considerably during erection without obstructing the lumen of the urethra. Only in this way a normal sperm transport during ejaculation can be guaranteed for. Skin from preputial origin seems to fulfil all these criteria and is therefore used as a direct replacement of the urethra. Firstly described by van Hook (1896) the use of the prepuce became more widely used after the publications of Asopa et al. (1971), Standoli (1979) and Duckett (1980). These authors describe the use of a pedicled flap to reconstruct the neo-urethra. Hagerty and Taber (1958) were the first who studied in more detail the bloodsupply of the different penile tissues. They performed a perfusion of the iliac and femoral vessels of stillborns with plastic material. After digestion of the surrounding tissue they found that the bloodsupply of the penile skin including the prepuce was supported mainly by the external

puddental artery.

The elaborate animal studies in dogs by Ninomiya and Nakamura (1981) showed in detail the vascular architecture of the canine prepuce. The prepuce receives its bloodsupply for the outer layer mainly of an artery (dorsal/ventral preputial artery) originating from one of the branches (dorsally and ventrally located) of the caudal superficial epigastric artery that is one of the terminal branches of the external iliac artery. This preputial artery gives than branches (the so called recurrent arteries) to the inner prepuce, communicating at the preputial fornix with branches of the deep dorsal penile artery, that originates from the internal iliac artery. So the bloodsupply of the prepuce in dogs is mainly dependant on the external iliac artery with many communications through the recurrent arteries with the internal iliac artery (fig. 6).

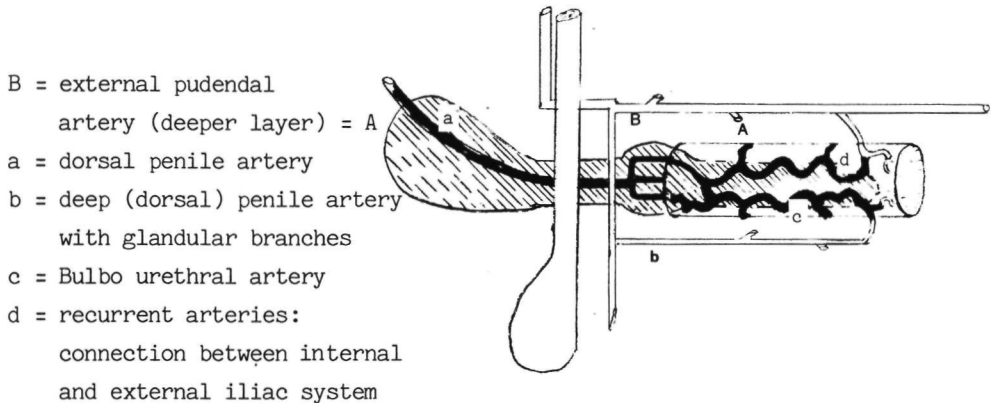


Fig. 6: The bloodsupply of the canine prepuce
(schematically after Ninomiya and Nakamura, 1981)

This longitudinally orientated vascular pattern allows for the total preparation of the penile skin including the prepuce as an independantly vascularized flap, with separate vascular support of the two preputial layers. Our investigations, schematically represented in fig. 6, showed that a quite similar vascularisation pattern exists in the human penis with separate bloodsupply of the penile body and the penile skin including the prepuce.

The external pudendal artery divides at the base of the penis (comparable with the fornix of the canine prepuce) in two major longitudinally orientated systems: the superficial branches supporting the penile skin and outer prepuce and the deeper branches, supporting the inner prepuce. This vascular architecture allows the dissection of an independently vascularized pedicle of the inner prepuce leaving the penile skin bloodsupply undisturbed. Thanks to the large number of communicating vessels between the inner and outer prepuce also a double faced island flap using the inner prepuce for the reconstruction of the neo-urethra and the outer prepuce as it skin cover, can be prepared. The bloodsupply of the outer prepuce depends in these cases on the deeper vascularization by the external pudendal artery.

Results of hypospadias correction with the transverse inner preputial island flap technique.

5.1 Introduction

This study compares retrospectively the results of the transverse inner preputial island flap technique for hypospadias repair with the results of other large series from the literature.

5.2 Patients and methods

5.2.1 Patients

From May 1979 through April 1984 a total number of 142 patients were treated for hypospadias at the department of urology of the Academic Hospital of the Rijksuniversiteit and at the department of urology of the Maria Middelaers Hospital, both in Ghent, Belgium.

In the period from August 1979 until November 1984 a total number of 87 patients were treated for hypospadias at the department of urology of the St. Radboud hospital of the Catholic University in Nymegen, the Netherlands.

To be eligible for this study the following criteria had to be met by the patients:

a correction with the transverse inner preputial island flap technique (119 from 229 patients).

- b a follow up of at least one year (115 from 119 patients). The follow up varied from 12 to 42 months, with a mean of 16 months. In Belgium the mean follow up time was 19, in the Netherlands 14 months.
- c last out-patient control should have been less than six months (Belgium); or less than two months before the end of this study (the Netherlands).

According to these criteria 61 out of 142 patients in Belgium and 54 out of 87 patients of the Dutch group were eligible for entry into this study.

In Belgium, 81 patients with hypospadias correction were not included in this study for the following reasons:

- a. another type of operation: 78 patients
- b. follow up less than one year: 3 patients

In the Netherlands, 33 patients were not eligible for this study for the following reasons:

- a. another type of operation: 32 patients
- b. follow up less than one year: 1 patient

In most cases (n=110) another type of hypospadias repair was performed because of the minor degree of the malformation.

Age distribution: The age of the patients at the time of operation was between 1 2/12 and 22 years. The patients were divided into five age groups (fig. 1).

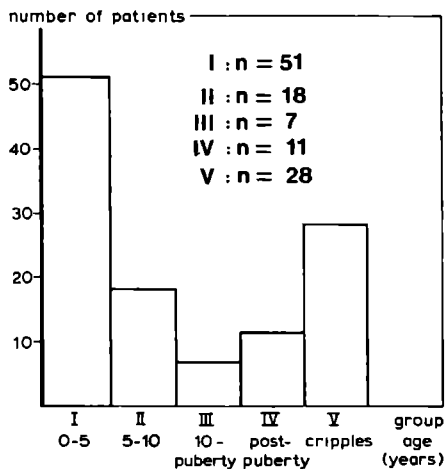


Fig. 1: Age distribution. Number of patients 115.

This classification in these 5 different groups has been chosen to be able to evaluate eventual age-related problems.

The patients in group I, 0 till 5 years, will have fecal incontinence with special post operative nursing problems.

In patients in group II, 5 till 10 years, this is in general no problem. Nocturnal detumescence will seldomly occur.

The patients in group III, 10 years of age until puberty, will in general have more nocturnal erections but the penis still has prepubertal dimensions.

The patients in group IV are grown up young men with frequent nocturnal erections and adult penile size.

In group V all the patients are included regardless of age which had at least two previous attempts of hypospadias repair (2 up to 8, mean 3.3 operations besides a meatotomy).

This is the so called "hypospadias cripple group". The four other groups together were called the "primary case group".

Severity of the hypospadiac anomaly: As explained before (chapter II) we prefer to classify our patients according to the two main features of the hypospadiac entity: the ectopy of the orifice and the bending of the penis (Chapter II, Table II). The ectopy of the orifice was graded from 0 to 10, in which grade 0 corresponds with a normal position, while grade 10 represents a maximal displacement.

In the same way the several degrees of chordee were numbered from 0 to 10. Absence of chordee was graded as 0. The mild degree (till 15 degrees deviation) is numbered from 1-3 (one number for every 5 degrees of deviation). The moderate chordee (between 15 and 35 degrees) is numbered from 4-7 (one number for every 5 degrees of deviation). The severe chordee (over 35 degrees) is numbered from 8-10 (one number for every 5 degrees of deviation). Combining these two elements a proper description can be given about the severity of the hypospadias.

If the sum of the numbers of the two characteristics is 5 or less than we speak of mild hypospadias, between 6 and 10 of moderate hypospadias and over 10 of severe hypospadias (fig. 2).

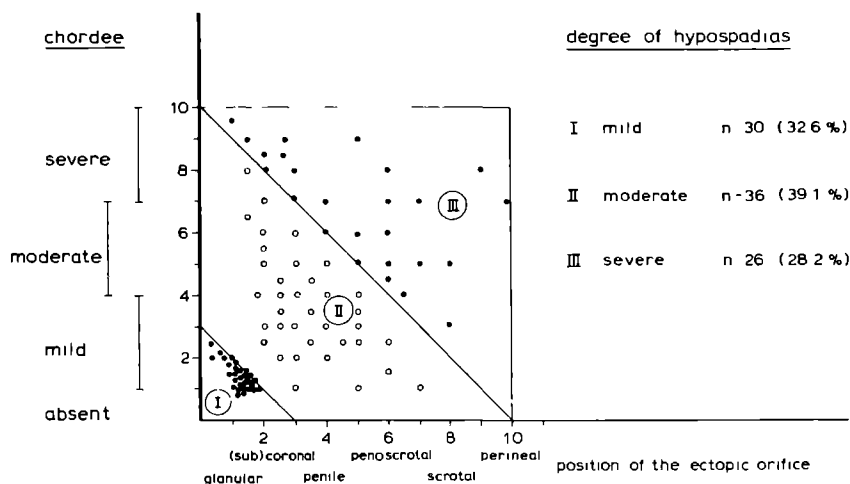


Fig. 2: Classification of the patients according to the severity of their hypospadiac anomaly: mild, moderate, severe. Total number of patients 92 (87 primary cases and 5 patients out of the "cripple group" with reliable preoperative data.

In our material there was one case in which the penile size before operation was less than 2 1/2 standard deviation of the normal penile size curve, while the other patients had a penile size within the normal limits (fig. 3).

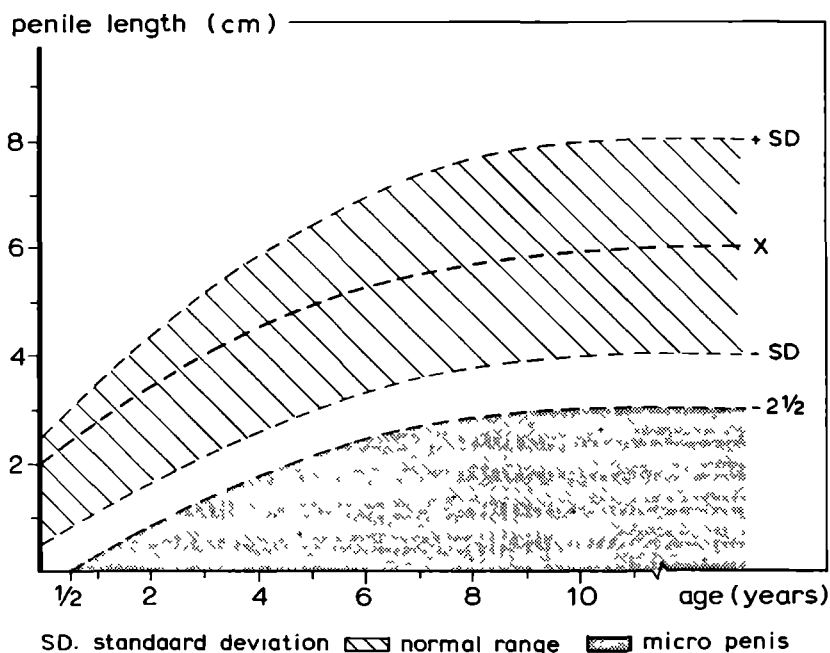


Fig. 3: Normal penile growth
Adapted from Stephens (1983)

5.2.2 Methods

The surgical procedure

The surgical procedure used for the correction of the hypospadias was the transverse inner preputial island flap technique, which is described in detail in chapter III.

This standard procedure was performed in 107 out of the studied 115 cases. In one case an additional interposition procedure (Devine and Horton 1977) and in 7 cases a dorsal supplementary correction (Nesbit 1961) had to be done. There were no age-related differences in the operative procedure. There was however a major difference between the group that was operated for the first time, the primary group (group I - IV, 87 patients) and the "cripple" group (group V, 28 patients). In the latter the

chordectomy was more complicated and an additional procedure was necessary to release the chordee completely in 4 out of 28 cases. Furthermore in patients from group V there was always a problem to cover the penis with skin. In most cases (8 out of 10 in the Dutch group), at least partly, penile skin had to be taken to make a vascularized tube for the urethroplasty, since there was not sufficient prepuce left after the previous operations. That this is feasible has been shown before (de Vries and Debruyne 1984).

Peri-operative management

There was a difference between the Belgian and Dutch group of patients in controlling the peri-operative loss of blood. In the Belgian group vasoconstrictive agents were used (de Sy et al. 1981). Personally we prefer the careful use of bi-and/or unipolar electrocoagulation without the use of vasoconstrictive agents. Also the policy for the administration of antibiotics was different in the Belgian and Dutch group. In the latter group no routine antibiotics were given peri-operatively (as done in the Belgian group), medication was administered only if infection was present and the result of the antibiogram was known.

Post-operative evaluation

The post-operative evaluation was based on the data that were collected from three sources:

- 1 The answers from the questionnaire
- 2 The hospital files
- 3 Physical examination of the patients

The questionnaire was send to the whole group of 115 patients. An abstract of the questionnaire is shown in Table I.

Table I: Abstract of the questionnaire

Family history:

Fathers name, date of birth,

previous operations of the urogenital tract.

Mothers name, date of birth, medication during pregnancy,
duration of pregnancy.

Other malformations known in the family.

Patients history:

Birth weight, all previous operations, other abnormalities,
age at first corrective operation for the hypospadiac
malformation, duration of hospital stay (if more than one
the number of days of each operative procedure).

Post operative complaints.

General remarks:

Grade of satisfaction with the overall results.

Comments about the age at which the operation was performed.

Comment about the sexual functions (in appropriate cases).

Other remarks:

The response rate was 100 % in the Dutch group and 83.6 % in the Belgian group, which represents an overall response rate of 92 %. From ten patients it was not possible to get complete information for different reasons (lost from follow-up; incomplete information).

Besides relevant epidemiological data this questionnaire provided us with the data concerning the subjective results of the transverse inner preputial island flap repair.

Information on the peri-operative management and the duration of the admission to the hospital were obtained from the patient files.

A physical examination of the patients was performed on an outpatient base in all patients less than six month or two month in respectively the Belgian and Dutch group before the end of the

study. At this occasion we also tested the sensibility of the penis after this extensive corrective procedure. For this purpose a sensitivity test was developed.

The investigation was performed in such a way that the patient was not able to observe the actual testing procedure. After the glans was cleaned, it was touched with a blunt needle as used in a routine neurological investigation. The glans was divided into four segments with the neo-meatus in the center. Starting at twelve o'clock the four segments were numbered one to four in a clockwise direction. In each segment a total number of ten tests were performed. The at random chosen area of the glans was touched with the needle with a pressure of approximately 5 grams and the patients were asked to indicate whether or not the penis was touched and if so in which segment. A segment in which the number of correct answers was less than six, was considered to have lost sensibility.

An other important factor for the final judgement about the result of the reconstruction was the possibility of the patient to empty his bladder completely within normal time limits. To measure this characteristic a flowmetric investigation was performed. We used a standardized mictrometer (Disa Elektroniks) providing a flow time curve during micturation (Rollemma 1981). A flowrate of 10 ml/sec in patients before puberty and of 14 ml/sec after puberty were regarded as lowest normal value. In children under the age of five this test usually can not be performed and an actual measurement with a bougie à boule was performed. A diameter of both the meatus and the proximal anastomosis of 10 charriere was accepted as normal.

The final outcome of the operation was categorized into three groups. The standard to which we compared our results with is the patient who had in his early childhood an uneventfully healed circumcision. The final result of the corrective procedure is categorized as:

Good:

if the penis of the patient appears to be simply circumcised and a normal micturation with a normal flowrate can be established.

Fair:

if there is a minor difference, from an aesthetical point of view, with the circumcised penis but with a normal function. Neither the patient nor the surgeon feel the urge for additional operative correction.

Bad:

all those situations in which at the end of this study the functional aspects (diminished flowrate, fistula and/or rest-chordee) needed further operative correction, or in which the outlook was not acceptable compared to our standard, with or without further functional limitations.

5.3 Results

5.3.1 Epidemiological data

Maternal factors:

The age of the mothers of the 105 hypospadiac boys was between 18 1/2 years and 38 4/12 years, mean 28 2/12 year.

During pregnancy medication was taken by 9 out of 105 mothers for urinary tract infections. In the Belgian group, 2 women received Human gonadotrophine, in one case there was some bleeding during the first trimester, the other woman was known to have subfertility problems. Four other women received progesterone derivatives for bleeding periods during the first trimester of the pregnancy. One woman needed antidiabetic medication. Two mothers used mild hypotensive drugs.

In our series there were 8 children premature and 5 out of this group were dysmature.

Heridity

In 18 out of 105 patients (17.2 %) one or more members of the family were affected with hypospadias (Table II). Because we are not sure that the total number of relatives was adequately present in our series (only 60 % of the questionnaires was confirmative) we do not dare to give an incidence for the whole sibship. The total number of affected relatives is 39 in our series.

Table II: Familial cases of hypospadias.

Affected family members	number of families

Index case only	87
+ father	1
+ father and another brother	2
+ brother	5
+ cousins	10

Associated anomalies

In our group a bilateral maldescensus was present in 11 cases and a bilateral cryptorchidism in one case. In eight cases there was a unilateral maldescensus (Table III). This represents an overall incidence of 17 % .

Table III: Maldescensus testis in relation with the severity of hypospadias.*

Chordee	Position of the orifice				
	distal	penile	penoscrotal	scrotal	perineal
absent	-	-	-	-	-
mild	-	I, 1	II, 2	-	2
moderate	-	II	II	-	1
severe	-	-	II, 1	III	1
				(one with cryptorchidism)	

* Roman and arabic figures indicate respectively bilateral and unilateral maldescensus testis.

Besides maldescensus testis urogenital and other anomalies were found in 29 patients. In 9 cases these anomalies were accompanied by maldescensus (Table IV).

Table IV: Anomalies other than maldescensus testis associated
with hypospadias

Urogenital anomalies:

Urethral obstruction	2
Vesico-renal reflux	1*
Bilateral renal hypoplasia	1
Duplex system	1
Utricle cyst	8 (2* 1+)
Wilm's tumor	1 +

Genetic defects

Deletion of long arm Y chromosome	1*
XY/XO	1
20-22 Desmolase deficiency	1*
DHT deficiency	1*

Other anomalies

Congenital heart defects	5 (2*)
Strabismus	4 (1*)
Congenital glaucoma	1
Pylorus hypertrophy	1

* combined with maldescensus testis

+ patient with Wiedemann Beckwith syndrome

From the 14 urogenital anomalies (13 patients), only 5 of these were serious enough to need surgical correction.

5.3.2 Appreciation of the result by the patient

The appreciation of the results is based upon both aesthetical and functional aspects. Both these aspects can be appreciated differently by the surgeon and the patient.

The questionnaire answers gave a good insight in the appreciation of the results by the patients or their parents. In 108 cases a definite judgement of the final results of the operation was

given (Table V). In 87 cases (80.6 %) this final judgement expressed satisfaction with both the aesthetical and functional aspects. In 11 cases (10.2 %) the patients were just satisfied with the result with minor complaints about aesthetical aspects (9 cases) or about the size of the penis (2 cases). None of these patients expressed the wish for further corrections. In 10 cases (9.2 %) the patients or their parents were disappointed about the final result. In 5 cases there still existed a fistula (all cases closed successfully after termination of this study). In 4 cases the cosmetic appearance of the penis was found disappointing. In one case scars not related to the operation but the result of severe burning due to a faulty earth connection of the electro coagulator were the reason for a negative judgement.

Table V: Appreciation of the hypospadias correction
by the patient

	number of patients	percentage
Good	87/108	80.6 %
Reasonable	11/108	10.2 %
Bad	10/108	9.2 %

5.3.3 Sensibility of the glans after operation

A total number of 25 patients were investigated postoperatively for the sensitivity of the glans. (16 out of the primary case group, 9 out of the "cripple" case group with reliable preoperative data). In all these patients there existed according to the patient a normal sensibility of the glans preoperatively. The sensibility was completely intact in 14 out of 16 primary cases: 87.5 % and in 3 out of 9 "cripple" cases: 33.3 %. In the primary case group sensibility loss was restricted to one segment in 1 patient and to two segments in another patient. In the "cripple" group sensibility loss was restricted to one segment in

1 patient, to two segments in 3 patients and to three segments in another 2 patients (Table VI).

Table VI: Results of the sensibility test

Sensibility	Primary case group*		"Cripple" case group**	
	normal	absent	normal	absent
segment				
I dorsal left	16	-	8	1
II ventral left	15	1	3	6
III ventral right	14	2	4	5
IV dorsal right	16	-	8	1

* 16 patients out of the primary case group

** 9 patients out of the "cripple" case group

All patients over 10 years of age

In this study 10 patients were able to distinguish between the meatal part that was formed out of the inner preputial island flap and the adjacent glandular tissue.

5.3.4 Evaluation of functional aspects of the neo-urethra

Of all patients postoperative data were available. They were based on either flowmetric investigations (n=35), actual bougie à boule measurements (n=50) or combined investigations (n=30). Of all flowmetric measurements only 2 patients showed abnormal flow-time curves and a stenosis was subsequently diagnosed. They were counted as a bad result (group of primary cases). The bougie à boule measurement showed in five cases a possible obstruction, in 2 patients at the proximal anastomosis and in 3 at the meatus (n=3). Further investigations showed in 2 cases a real obstruction (both with fistula formation) and were counted for as bad result ("hypospadias cripple" group). In the remaining group of 30 patients absolutely no sign of any obstructive pathology

was established and the micturation was considered to be normal.

5.3.5 Final results

The results did not show a significant difference between the different age groups. A good result was obtained in group I, II, III and IV, in respectively 41 out of 51 (80.3 %), 14 out of 18 (77.8 %), 5 out of 7 (71.4 %) and 8 out of 11 patients (72.7 %). There existed however a significant difference between the "primary case" group (Group I, II, III and IV) and the "hypospadias cripple" group (Group V). For this reason the results will be presented for these two groups only. The final result from both functional and cosmetic point of view was considered to be good in 68 out of 87 primary cases (78.2 %) fair in 14 out of 87 primary cases (16.1 %) and bad in 5 out of 87 primary cases (5.7 %) (fig. 4).

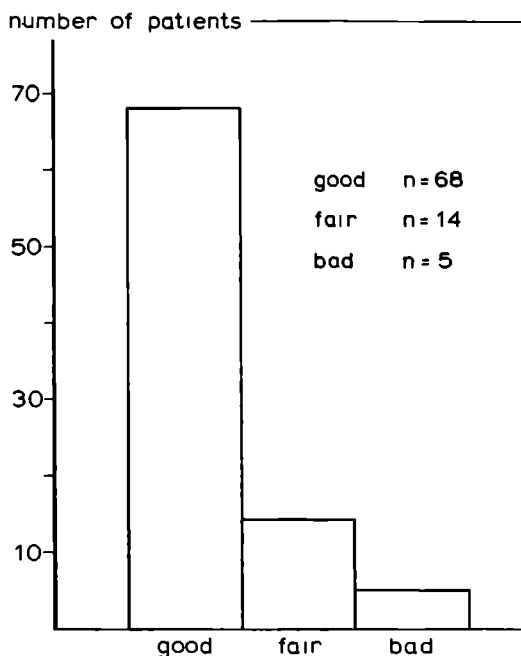


Fig. 4: The final result after transverse inner preputial island flap hypospadias repair in 87 primary cases

Patients with a good or fair result did not need further operative corrections. In the group of patients with a bad result (n=5), in two cases an obstructive process at the meatus has diagnosed, but surgical correction has not been performed yet. In one of these cases the parents refused a re-operation, because they were satisfied with the obtained result. In the other case the micturation is felt to be undisturbed by the patient himself. In three cases the cosmetic aspect of the penis is not good, but no further operations are planned.

In the primary case group 121 sessions under general anaesthesia (dilatations or operative procedures) were necessary in 87 patients (fig. 5). There was a major difference between the Belgian and Dutch group of patients in the number of sessions (operations), that was needed to achieve these results. In the Belgian group 1.2 operations per patient were necessary (53

operations in 43 patients) while in the Dutch group 1.5 operations per patient were performed (68 operations in 44 patients).

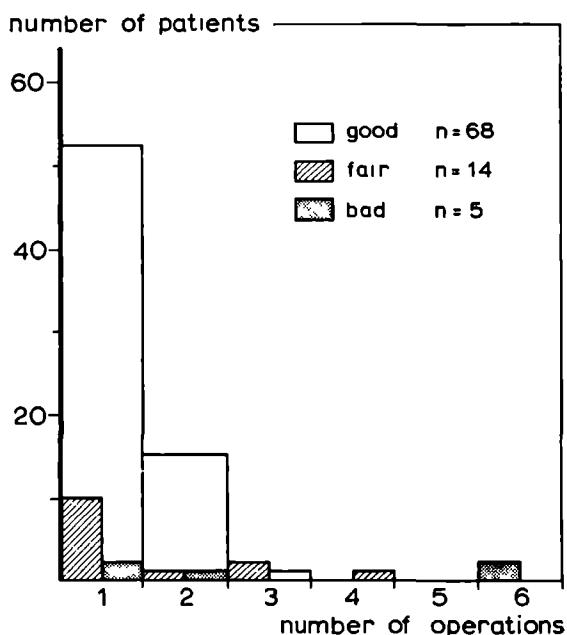


Fig. 5: Results in relation to the number of operations that was performed in the primary case group (n=87)

The main reason for this difference can be found in the growing experience with this type of operation, that is necessary to avoid complications (fig. 6).

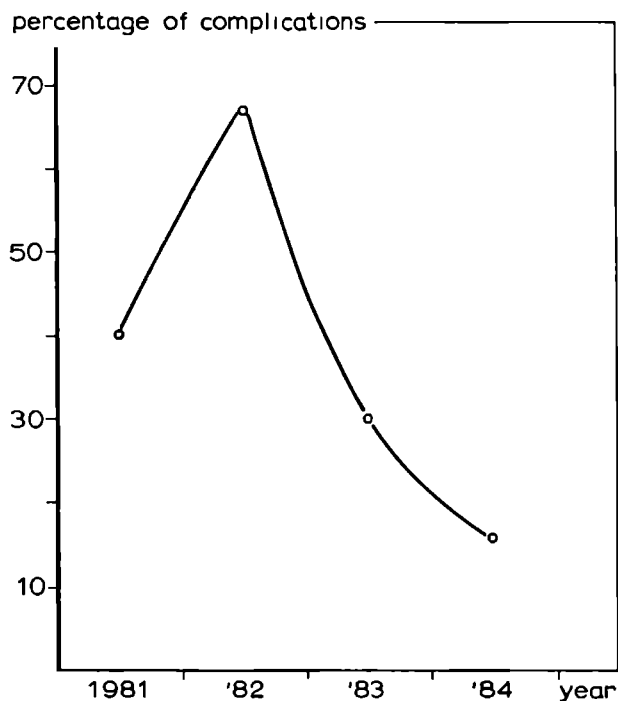
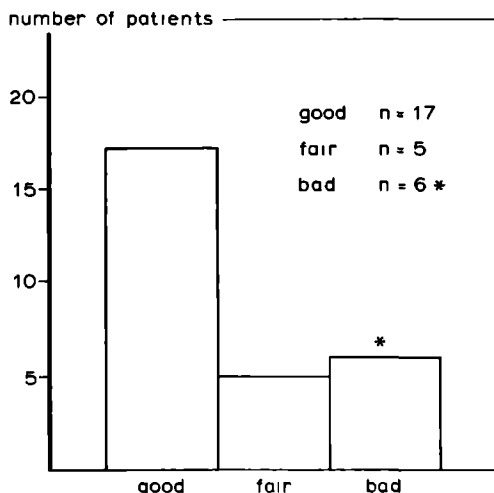


Fig. 6: The learning curve: percentage of complications in relation to growing experience

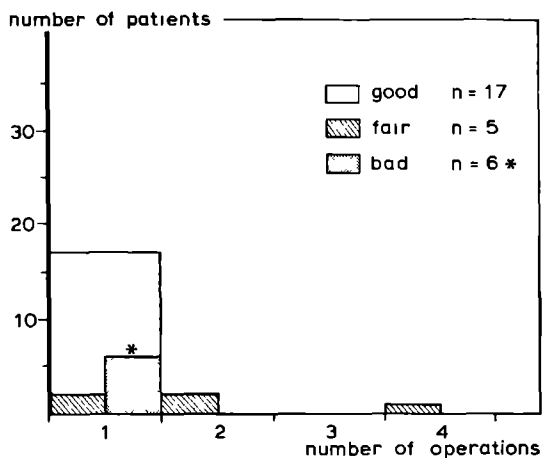
The final result in the "hypospadias cripple" group was considered to be good in 17 out of 28 "hypospadias cripple" cases: 60.7 %, fair in 5 out of 28 "hypospadias cripple" cases: 17.8 %, bad in 6 out of 28 "hypospadias cripple"cases: 21.4 % (fig. 7).



* Fistula in 5 cases: all successfully closed after the time period of this study (good result in 4 cases) fair result in 1 case

Fig. 7: The final result after transverse inner preputial island flap hypospadias repair in the "hypospadias cripple" group (n=28)

In the group of patients with a good or fair result no further corrections are anticipated. In the group of patients with a bad result (n=6) one patient had such ugly burning scars related to the electro cautery, that the result was called bad, however the functional and cosmetic aspects of the hypospadias repair were fair. In 5 patients there exists still a fistula at the termination of this study. Four of these patients have been operated successfully within half a year after this study and one is closed recently. These results were achieved in 33 operative sessions in 28 patients. The ratio between the Belgian and Dutch groups were 18 operations in 18 patients and 15 operations in 10 patients respectively (fig. 8).



* Fistula in 5 cases all successfully closed after the time period of this study (good result in 4 cases) fair result in 1 case

Fig. 8: Results in relation to the number of operations that was performed in the "hypospadias cripple" group (n=28)

5.3.6 Complications

The complication rate in the separate age groups I, II, III and IV was very similar (Table VII).

Table VII: Complication rate per group

Group I	39.2 % (20 out of 51 patients)
Group II	38.9 % (7 out of 18 patients)
Group III	42.9 % (3 out of 7 patients)
Group IV	36.4 % (4 out of 11 patients)

Therefore these groups are considered together as one group: the

primary case group with a total number of 87 patients.

Per-operative complications

During surgery no major complications, like severe bleeding, anaesthesiological problems, or damage to neighbouring organs, did occur. Furthermore, no problems were encountered during surgery which made a change of the planned procedure necessary. There was no mortality. One major complication was caused by a faulty connection in the coagulating equipment. Because of a defective security module the equipment was not automatically turned off. This caused severe second and partially third degree burning wounds on the buttocks. The hospital stay was prolonged for three weeks and the boy was dismissed with well corrected external genitalia but with ugly scars (patient of Group V).

Early post-operative complications

In the immediate post operative period bladder spasms were the most frequently observed problem. They were treated successfully with anticholinergic and/or spasmolytic agents. In eight cases we observed superficial necrosis of the skin, which was always limited to the edges of the penile skin flaps. In two cases a more extensive dehiscence of the wound was seen, not directly related to infection. Infection also formed a major complication. The more superficial infections never formed an indication for surgical intervention. The more severe forms of infection were encountered both with and without the use of routine antibiotics. These more serious infections led to the formation of a fistula in three patients and to wound dehiscences in two cases. These complications needed secondary repair. Post-operative bleeding was seen in six cases which made reintervention necessary in two patients.

Catheter problems were encountered in three cases. One unintended catheter (hooked up) removal on the first operative day caused a fistula. Catheter blockage was treated conservatively in two cases and we had to replace it under general anaesthesia in one

patient.

Edema never made re-intervention necessary (Table VIII).

Table VIII: Early post operative complications
(all patients)

complication	number of patients	outcome
Haemorrhage	6 (1)*	2 (1) reinterventions 1 fistula
Edema	4	
Infection superficial	5 (3)	
severe	5 (3)	3 (2) fistula and 2 (1) dehiscence
Dehiscence	2 (1)	2 (1) fistula
Necrosis superficial	8 (5)	
urethra	-	
Catheter blockage	2 (1)	1 fistula
accident	1	
Severe bladder spasm	9 (1)	

* The figures in parenthesis are complications observed in the "cripple" case group (n=28).

Late post-operative complications

The two major late post-operative complications were fistula and stricture formation at one or both anastomoses of the neo-urethra (Table IX).

Table IX: Late post-operative complications
(all patients)

complication	number of patients	outcome
Fistula	19 (8)*	5 persistent fistulas
Stricture formation		
meatal	9 (3)	2 subnormal flow
proximal	4 (1)	
Chordee	2 (1)	
Torsion	3	
Bladderstones	2	
Skin de-arrangement	1	

Patients with both stricture and fistula formation are reported in the fistula and in the stricture group.

* The figures in parenthesis are complications observed in the "hypospadias cripple" group (n=28).

Although most of the fistulas are already seen in the immediate post-operative period, they are usually not repaired immediately. Persistent fistula formation (seven fistulas closed spontaneously) was seen in 11 out of 87 patients of the primary group (12.6 %). In the "hypospadias cripple" group (n=28) 8 persistent fistula were seen (28.6 %).

The recurrence rate of the fistulas after the first attempt to close them was 3 out of 11 in the primary case group and 1 out of 4 in the "hypospadias cripple" group (5 fistulas were closed after the end of this study).

The most-annoying post-operative complication is the formation of a stricture especially at the meatus. This occurred in 9 out of 87 cases (10.3 %) in the primary case group and in 4 out of 28 cases (14.3 %) in the "hypospadias cripple" group. In 11 patients this complication was accompanied with fistula formation. In our earlier material, this complication alone was responsible for 11 consecutive procedures in two children, both with fistula formation.

In three patients a torsion of the penis was found, which made a secondary repair necessary in one case.

There were two cases with persistent chordee, which was of such a minor degree, that neither the surgeon nor the patient felt correction necessary.

The two patients with bladderstones had, beside hypospadias, severe other anomalies, so that prolonged catheter drainage was necessary.

One patient had such an asymmetrical arrangement of skin, that correction had to be performed together with a minor repair at the meatus because of stricture formation.

5.4 Discussion

5.4.1 Indications for surgery

The need for operative correction of moderate and severe forms of hypospadias, with a meatal dystopy and bending of the penis, is obvious. There is still debate whether milder forms also need surgical correction. Many authors suggest for this latter group a thoughtful resignation (Cabot 1936, Heybroek 1964, Culp and Roberts 1968). Nowadays however no surgeon can deny the psychological sequelae of even mild hypospadias (Kenawi 1976, Avellán 1976, Ehrhardt 1979, Berg 1983). We must however bear in mind that; "Hypospadias can undoubtedly cause undesirable psychological reactions. Yet what are the psychological consequences for the patient that is forced to undergo an endless serie of "curative" operations?" (Hand 1948).

We think that the indication for operative correction of any hypospadiac malformation of any degree is given by the patient himself, or in the case of children, their parents.

5.4.2 Patients and methods

Due to the referral function of academic hospitals there exists in our patient population an overrepresentation of severe forms of hypospadias. This holds especially for those patients in group V who came for operative correction in the presence of fistulas and rest chordee.

This bias in selection of the patients should be taken into consideration, when the results are analyzed.

The classification of the hypospadiac entity in mild, moderate and severe forms by evaluating the two main characteristics of this malformation, enables a good, reproducible description. The use of this classification would make it possible to compare the results of different publications about this malformation.

Using this classification we found for reasons mentioned above a significantly higher incidence of 28.2 % (26 out of 92 cases, fig. 2) of severe hypospadias in our group, compared to the literature, in which an incidence of 8.7 % (256 out of 2948 cases) of severe cases is found (Chapter I, Table III).

A further explanation for the high incidence of severe forms in our material is the high frequency of children (5.2 %: 6 out of 115 patients) with concomitant chromosomal, enzymatic and metabolic disorders.

The mean age of the mother in our hypospadiac patients at delivery (28 3/12 years) is not significantly different from the mean age (27 3/12 years) of mothers who gave birth to healthy boys in the same period of time (Central Bureau of Statistics, the Netherlands, 1984).

Out of 105 mothers, 6 mothers (5.7 %) received hormonal treatment during early pregnancy. This incidence is too low to confirm the findings of Neto et al (1981) and Angerpointer (1984), who stated that this medication could be a causal factor for the hypospadiac entity.

The prematurity rate of 7.6 % in our group is within the normal limits of a large control population in which a rate of 10.6 %

was found for boys (Källén and Winberg 1982). The dysmaturity rate of 4.7 % in our group is not higher than the rate in the control group (4.2 %) in the above mentioned Swedish study.

The number of patients in our study with affected relatives (18 out of 105 cases) is in concordance with data from the literature (chapter I, Table VII). The total number of affected relatives in our patient group (31 out of 105 families) is in the same range (80 out of 240 families) as found by Bauer et al. (1979).

We found a positive correlation between the severity of the hypospadias and the incidence of maldescensus testis (chapter I, Table X). In the literature a correlation has been found for the incidence of maldescensus testis and the frequency and severity of the utriculus cyst formation (chapter I).

In our patient group we found in 29 out of 115 cases associated anomalies (excluding maldescensus testis). This incidence of 25.2 % is very high. To be able to compare it with the data from the literature we should subtract the 6 non-symptomatic utriculus-cysts since these anomalies are not mentioned, in literature. This correction would give an incidence of 20 % (23 out of 115 cases). Our higher incidence can be explained by the relatively large number of severe cases.

Although we found a correlation between the severity of hypospadias and the presence of Müllerian remnants and maldescensus testis, we did not observe a correlation with other associated anomalies, as has been stated in the literature (Svensson 1979, Khuri et al. 1981). However, other authors also could not confirm this correlation (Lutzker et al. 1977, Shima et al. 1979, Devine et al. 1980).

In our material we found in 7 out of 115 patients (6.1 %) 8 urological anomalies besides the maldescensus testis and asymptomatic utriculuscysts.

In the literature the incidence for associated urological anomalies (not including the maldescensus testis) varies considerably from 2.2 to 14 % . Svensson (1979) found 13 out of 84 investigated cases out of 625 hypospadiac cases. Shima et al. (1979) found 109 urogenital anomalies out of 272 cases of which almost 6 % needed surgical correction. Khuri et al. (1981) found 48 significant anomalies among 460 investigated patients, of which the majority needed surgery. Overall, besides hypospadias and maldescensus testis, one may find 4 to 5 % anomalies of the urogenital tract, that eventually need to be corrected. This is twice the incidence as found in non-hypospadiac males (Opitz 1985). Our incidence of urological anomalies of 6.1 % is in concordance with these data. All our patients had symptoms of their concomitant urological anomaly; in 6 cases recurrent urinary tract infections, in one case an abdominal mass (patient with a Wilms tumor) and in another case more general symptoms like growth retardation formed an indication for further urological investigation. In none of the hypospadiac patients without further symptoms an additional surgically correctable malformation was found during urological investigation. From these observations it is clear that hypospadiac patients without further symptoms do not need a full uro-radiological investigation. This confirms the findings of other authors (Kelly et al. 1984 Opitz 1985, Kroovand 1985, Kaplan 1985, and Cerasaro et al. 1986), that these investigations are not necessary if there exist no further symptoms.

5.4.3 Results

The results of this study are first compared with those reported in the literature also using a one-stage repair technique and secondly with some still widely used two- or more stage techniques.

First we will review the results of the hypospadias correction with other one stage techniques than the pedicled vascularized island flap technique (Table X, XI, XII and XIII) and there after

the results obtained by using the inner preputial island flap technique (Table XIV).

Table X: Results of one stage repair in mild hypospadias, not using island flap techniques*

Author	year	number of patients	Fistula and/or Dehiscence	Urethral stricture	Meatus Stenosis	Dystopy
Devine and Horton	1977	100	13 %	-	2 %	
Filmer et al.	1977	20	30 %	5 %	5 % (+Fistula)	
Shubailat and Ajluni	1978	62	7 %	-	2 %	
Kim and Hendren	1981	50	-	-	4 %	
Rees et al.	1981	33	21 %	-	N.D.**	
Belman	1982	30	10 %	-	10 %	
Bochiotti et al.	1982	21	15 %	-	N.D.**	
Woodard and Cleveland	1982	76	13 %	1 %	13 % (2 % with fist.	
Hanna	1983	21	5 %	-	5 %	
Redman	1983	60	28 %	7 %	21 %	

* Operative procedure: Horton and Devine (flip-flap) technique and variations

** N.D.: no data given

Table XI: Results of one-stage repair in mild hypospadias,
not using island flap techniques*

Author	year	number of patients	Fistula and/or Dehiscence	Urethral stricture	Meatus Stenosis	Dystopy	Chordee
Allen and Spence	1968	28	4 %	-	-		
Hinderer	1971	30	23 %	N.D.**	N.D.**		
Sadlowsky et al.	1974	85	3.5%	-	1 %		
Meyersfield et al.	1977	31	3 %	-	3 %		
Marshall et al.	1978	102	3 %	-	2 %		
Mills et al.	1981	19	5 %	-	5 %		
Gonzales et al.	1983	63	8 %	-	2 %		
Lima et al.	1983	74	8 %	-	2 %		

* Operative procedure: Bevan, Ombredanne, Mathieu or King technique

** N.D.: no data given

Table XII: Results of one-stage repair in mild and moderate hypospadias
not using island flap techniques*

Author	year	number of patients	Fistula and/or Dehiscence	Urethral stricture	Meatus Stenosis	Dystopy	persistent Chordee
Tolhurst and Gorter	1976	102	-	3 %	-		-
Henderson	1981	143	4 %	1 %	1 %		-
Perovic	1983	63	5 %	-	-		2 %

* Operative procedure: Denis Browne technique and variations.

Table XIII: Results of one-stage repair for moderate and severe hypospadias, not using island flap technique*

Author	year	number of patients	Fistula and/or Dehiscence	Urethral stricture	Meatus Stenosis	Dystopy	Persistent
Devine and Horton	1977	17	50 %	15 % (one with fistula)			
Filmer et al.	1977	20	25 %	5 %	5 %		
De Sy et al.	1981	27	15 %	4 %			
Woodard and Cleveland	1982	75	20 %	-		9 % (7 % with fistula)	
Hanna	1983	28	39 %	7 %		18 % (7 % with fistula)	

* Operative procedure: Josserrand technique of free grafts and variations

In the first group of patients (Table X) operated with the one-stage technique according to Devine-Horton the fistula rate varies considerably from zero to 30 % . Urethral strictures are seldomly seen but meatal stenosis occurs in all studies (in two studies no exact data are given).

If post-operatively an ectopic orifice is accepted (Table XI and XII) less problems with the neo-meatus are encountered. Furthermore these techniques have a low incidence of fistula formation. The majority of the cases described in table X, XI and XII are mild hypospadias. Overall in this group (1213 patients) the fistula rate of 9 % and the formation of urethral and meatal strictures with a frequency of 3 % are lower than the complication rate in our study.

One should realize however, that in our study a considerable part of our patients had a more severe form of hypospadias, in contrast to these studies. Furthermore the techniques listed in table X, XI and XII give a location of the neo-meatus at the ventral part of the penis. This will lead to a less physiological appearance of the penis compared to the outcome after operation with the technique that we used.

The high complication rate of 26 % fistulas and 10 % anastomosal problems in the group of patients operated with a free graft technique (Table XIII) reflects the severity of the hypospadias on one side and the inherent problems of a non-vascularized tube on the other side.

The results of the one stage repairs using a pedicled vascularized island flap are given in Table XIV.

Table XIV. Results of one stage repair for moderate and severe hypospadias, using island flap techniques*

Author	year	number of patients	Fistula and/or Dehiscence	Urethral stricture	Meatus Stenosis	Dystopy	Chordee
Broadbent et al.	1961	8	38 %	10 %			
DesPrez et al.	1961	10	33 %	10 % (dilatation only)	20 % (dilatation only)		
Toksu	1970	15	20 %				
Asopa et al.	1971	12	30 %				
Engel and Scott	1973	40	10 %	15 % (5 % with fistula)	8 %		
Hodgson	1975	100	5 %	1 %	4 %		
Filmer et al.	1977	9		2 %			
Kroovand and Perlmutter	1980	47	6 %	23 % (20 % simple dilatation)			4 %
De Sy et al.	1981	17	6 %	12 %			
Duckett	1981	100			10 %		
Rees et al.	1981	15	47 %				
Standoli	1982	753	6 %	about 30 % (one or two dilatations)			
Hanna	1983	15	6 %	6 %			
Harris	1984	50	6 %		6 %		
Ransley	1984	33	18 %	9 %			
de Vries and Debruyne	1984	35	17 %	4 %	10 % (9 % with fistulas).		

* Operative procedure Broadbent, DesPrez, Toksu, Asopa, Hodgson, Standoli or Duckett technique with the use of a pedicled vascularized island flap.

The frequency of complications of the pedicled, vascularized flap technique as reported by other authors (Table XIV) with a fistula rate of 8 % (98 out of 1259 patients) and obstructive anastomosal problems of 20 % (251 out of 1259 patients) (including mild forms, which can be corrected with one or more dilatations) is comparable with the frequency of complications in our study with a fistula rate of 12.6 % in the primary group and 28.6 % in the "cripple" group and stricture formation in 10.3 % in the primary group and 14.3 % in the "cripple" group. It has to be taken into consideration that any obstructive problem also those which only need dilatation is counted as a major complication in our study, whereas in the literature this is not always mentioned as a complication. Table XIV also reflects clearly the fact that the complication rate decreases with increasing number of patients operated.

In general it is difficult to categorize the hypospadiac patients reported in the literature into mild, moderate and severe forms. For all patients listed in table X - XIV the overall fistula rate is 9.3 % and stricture formation is seen in 11.3 %.

If we divide the total group according to the severity of hypospadias, 905 patients had mild hypospadias and 1734 had a more severe form. The frequency of fistula or stricture formation in these patients is shown in table XV.

Table XV: Overall results of one stage hypospadias repair

Degree of hypospadias	number of patients	Fistula and/or Dehiscence	Stricture*
mild	905	94 (10.4 %)	24 (2.7 %)
more severe	1734	151 (8.7 %)	274 (15.8 %)

Total number of patients: 2639

* Including the mild forms which needed one or more dilatations

This table clearly shows that frequency of fistula formation is

not different but that stricture formation is more frequently seen after correction of more severe forms. However, in our study the severity of the hypospadias formed no discriminative factor for the outcome of the operation, but the fact whether or not the patient had undergone previous operations.

With these described one stage techniques in every fifth patient a complication is encountered that made a reintervention necessary.

The disadvantages of the two or more staged hypospadias repairs are obvious.

Despite this, at this moment these methods are still widely used and a comparison of these techniques with the inner preputial island flap technique seems appropriate.

The rate of fistula and stricture formation for the meatal-based flap repairs for mild hypospadias are given in table XVI, while in table XVII and XVIII the results of this method for the repair of more severe forms of hypospadias are given.

Table XVI: Results of multi stage repairs for mild hypospadias*

Author	year	number of patients	Fistula and/or Dehiscence	Urethral stricture	Meatal Stenosis	Dystopy
Heybroek	1964	20	25 %	-	N.D.**	N.D.**
Sykes	1972	140	12 %	-	1 %	10 %
Barcat	1973	55	5 %	-	-	-
Fuqua	1973	54	2 %	-	-	-
Abramovic	1981	396	12 %	-	10 %	-
Hautmann	1981	32	3 %	-	7 %	-
Hendren	1981	140	2 %	2 %	3 %	-
Hensle and Mollit	1981	30	7 %	-	3 %	-
Gharib	1984	182	11 %	-	11 %	-

* Operative procedure: Ombredanne, Matthieu or Belt-Fuqua technique using meatal-based flaps

** N.D.: no data given

Table XVII: Results of multistage repair in moderate and severe hypospadias

Author	year	number of patients	Fistula and/or Dehiscence	Urethral stricture	Meatus Stenosis	Dystopy	Persistent Chordee
Browne	1953	121	7 %	N.D.			
Moore	1958	74	5 %	-			
Heybroek	1964	28	43 %	10 %		3 %	
Lamaker	1964	88	33 %	-			
Culp and Roberts	1968	318	14 %	-		17 % (meatus)	
Michalowski et al.	1970	459	30 %	1 %	1 %		12 pat. out of 15 after puberty
Heiss and Helmig	1974	127	27 %	N.D.		43 %	
Wray et al.	1976	647	19 %	-		-	
Kelalis et al.	1977	23	40 %	20 %		10 %	
Yarbrough and Johnston	1977	96	7 %	2 %	1 %		
Gearhart and Witherington	1979	64	17 %	6 %		3 % (meatus)	5 %
Bailen and Howerton	1980	27	5 %	-	10 %		
Blümel and Köhnlein	1980	44	27 %	2 %	5 %	7 %	
Johanson and Avellán	1980	286	4 %	-		great %	
Donnelly and Prenderville	1981	72	5 %	-	4 %		
Feiber	1981	44	56 %	15 %			
Fudicker and Albrecht	1981	20			7 %	25 %	
Marberger and Pauer	1981	183 (original and Duplay)	21 %	2 %	2.5 %	?	? (unsatisfactory 10 %)
Ringert et al.	1981	30	20 %	-		10 %	
Gharib	1984	123	22 %	2 %			

* Operative procedure: Denis Browne or Duplay II technique with a buried epithelial strip.

Table XVIII: Results of multistage repair in moderate and severe hypospadias

Author	year	number of patients	Fistula rate and/or wounddehiscence	Urethral stricture	Meatus Stenosis	Dystopy
Byars	1955	77	17 %	-	-	
Davis and Culp	1955	46	52 %	-	-	
Heybroek	1964	11	10 %	10 %	10 %	
Van der Meulen	1964	9	30 %	-	-	
Cronin et al.	1965	134	24 %	2 %		
Culp and Roberts	1968	224	11 %	15 %	15 %	10 %
Smith	1973	120	12 %	2 %	1 %	12 %
Wray et al.	1976	253	21 %	7 %		
Davis and Harrold	1980	32	30 %			
Novak et al.	1980	27	12 %	8 %		
Kelalis et al.	1981	135	3 %	3 %	4 %	
Marberger and Pauer	1981	130	15 %	2 %	2.5 %	
Rees et al.	1981	43	38 %	-	-	
Smith	1981	285	3 %	-	-	
Gharib	1984	147	16 %	3 %	4 %	

Operative procedure: Duplay I, Byar or Cecil technique.

Table XIX: Overall results of multi stage repair in mild, moderate and severe hypospadias

number of	Fistula and/or Dehiscence	Stenosis meatal or proximal
5744	17 %	(4634) 5 %

The overall rate of fistula formation of multistaged repair techniques as compiled from the literature on almost 6000 patients,

is 17 % with an overall rate of obstructive problems of 5 % (Table XIX). This means that at least one of every fifth patient need at least one operation more.

There are only a few authors that give the total number of operations needed for the correction of this complications. Deducting from their information about 20 % of the fistula and/or stenosis corrections fail and need one or two supplementary operative procedures.

Evaluation of the number of patients with the combined complication (fistula and stenosis) was not possible.

In these two or more stage procedures all the patients were at least operated upon twice and every fifth patient needed another (third) session with a 20 % change for yet another (fourth) session.

A less ideal, mostly subcoronal position of the neo-meatus, which tends to retract even to a more proximal position later on (up to 30 % of the cases) will occur in all these cases (Heiss and Helmig 1975, Berg 1983). Compared with our results in which in 61 out of 87 patients (70.1 %) a good or fair result could be obtained in one operative session in the primary case group, with the meatus at its physiological position, the results of the multi-staged repairs are unfavourable.

The results of our study in primary cases are comparable to the above mentioned results as reported in literature for the one stage repairs (Table XX).

Table XX: Overall results of the transverse inner preputial
island flap technique

	Number of patients	Results		
		good	fair	bad
primary case group	87	68	14	5
hypospadias "cripple" group	28	17	6	6

The hypospadiac "cripple" group is a totally different entity, presenting all grades of persisting chordee and pre-existent fistula with clearly more unfavourable results.

The mere fact that these results were achieved by two different surgeons demonstrates the reproducibility of the method. From the patient's point of view, the only interesting question will be how many operations are needed to obtain a good or fair result.

From the learning curve it is obvious that a surgeon has to operate enough patients in order to gain and maintain sufficient experience and to avoid unnecessary complications.

We think that ultimately the incidence of major complications with this operative procedure can be reduced to 15 % .

This operative method allows a single stage repair of any degree of the hypospadiac deformity at an early age with the creation of a penis that resembles the penis of a boy who underwent just a circumcision (fig. 9).

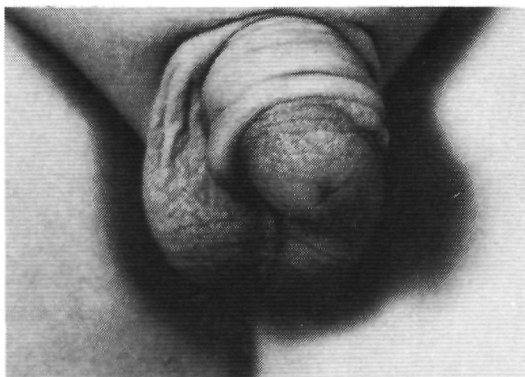


Fig. 9: The result after the transverse inner preputial island flap hypospadias repair in a moderate (left) and severe (right) hypospadiac case.

General Discussion

Introduction

The growing awareness of the "normal" outlook of the male genitalia in our more open society will possibly increase the number of patients with congenital malformations of the external genitalia which want corrective surgery.

The number of patients for this type of corrective surgery is further increased by the fact that repeated physical examinations at infant welfare centers will increase the number of referrals because of hypospadias.

The psycho-sexual consequences of the hypospadiac malformation have been recognised long ago (Zurhelle 1913). The interest in it was revived again by the study of Masters and Johnson (1966). Longterm follow up studies concerning sexual and psychosocial behaviour of hypospadiac patients became available recently. The study of Blotcky et al. (1978) showed a major difference in sexual behaviour between children operated on their genital tract and those with a non-genital operation (ear or throat). Robertson and Walker (1975), Lepore and Kessler (1979) showed the traumatic effect for the affected child of the awareness of his parents anxieties about the future development of the genitalia and particularly about the sexual function.

Another element that forms a severe burden for a normal psycho-sexual development is the secrecy about this "disease".

Heiss and Helmig (1975) showed that the major concern of the parents was not the ectopy of the meatus, but the presence of "dogears" or asymmetrical skin depots together with the bending of the organ at erection. For this reason correction of the chordee seems to be of utmost importance. This forms in our opinion a major drawback of two-stage procedures since a considerable number of the patients operated with these procedures

show a rest-chordee. Farkas (1970) found an incidence of rest-chordee in 17% while Heiss and Helmig (1974) reported even an incidence of 33 %. Avellan (1976) showed in a large review, that the development of puberty, the sexual debut and the sexual function differed in the hypospadiac cases from that of the normal Swedish male population. He found that the eagerness to undertake sexual activities in hypospadiac patients was less than normal. Berg (1983) states in his elaborate thesis on this subject that hypospadiac patients have a lower level of sexual activity and more dramatically even, a less qualified professional life relative to their objective qualities.

Age at operation

With this kind of operation we should try to prevent severe psychological and sexual disturbances in our hypospadiac patients. This justifies operation at an early age, before the identification with his sexual organ (so before the onset of speech and before development of the gender identity), has taken place (Ehrhardt 1979).

This limits the time at which the operation should be done from the newborn period to about the age of three years. In this period there are however several major psychological developments that should not be disturbed, unless there are compelling reasons to do so. The whole period is characterised by the Attachment-Separation-Loss feelings (Bowlby 1980). In this period the first months are vital for the mother-child relation and should not be disturbed. At the age of about fourteen to 24 months a period of Reapprochement of the child to the mother can be observed. So the most optimal age is around one year (Schultz et al. 1983). In our experience this age is also the most optimal, moreover since surgically there are no contra indications for operation at this age (Manley et al. 1981, Belman and Kass 1982). The smaller size of the penis at this age does not form a reason to postpone the operation, since the operative procedure is not hampered by this fact. In the questionnaire 40 % of the parents indicated that they would have preferred an operation at

an earlier age. This was mainly expressed by parents of children in group II (over 60 % of the parents). Only two parents of children out of group I would have preferred an operation at an older age.

Size of the penis in hypospadiac patients

In the literature there exists a very great variation about the size of the penis in hypospadiac patients. Farkas et al. (1969) found in their survey in 42 % of the cases a hypoplastic penis. Ericsson (1971) reported a hypoplasia of the penis of 20 % before surgical correction. Avellan (1976) described only 2 patients out of a total of 220 cases with hypoplasia, Kenawi (1976) described an incidence of 3.7 % while Heiss and Helmig (1975) just indicate in their large survey, that the average length was below the normal range (much more for the Browne as for the Ombredanne method). It seems likely that most of these differences can be accounted for by the scrutiny of the surgeon during the chordectomy and by the fact that hypoplasia of the penis is not standardized. In our opinion it should only be used if the size is not within the mean size ± 2.5 SD (Chapter V, fig. 3).

Using this criterium we had only one hypoplastic penis in our material.

Especially in the case of a hypoplastic penis problems can rise concerning the sex determination. The utmost endeavour should be undertaken to reconstruct the external genitalia according to the genetic sex. We think that, there is absolutely no reason to delay this kind of corrective surgery beyond the age of one year as we have been able to demonstrate in several of our severe hypospadiac cases. What traumatic consequences a wrong sex assignment can have, has already adequately been described in early literature (Fribe 1756, Cornelisse 1846, Neugebauer 1902, Zuhelle 1913). The appreciation of the operative results

There is a striking difference in certain follow-up studies of hypospadias correction with regard to the appreciation of the results by the surgeon or the patient himself.

In their material, Heiss and Helmig (1975) found that over 76% of the patients were satisfied with the operative outcome, however in over 40 % there were still anatomical abnormalities. Kenawi (1976) reported that 11 out of 182 patients were not satisfied with the appearance of their penis, of which five were reoperated for rest-chordee. In our own material, 105 (from 115) patients or their parents gave their appreciation concerning the final result. Just over 80 % were very satisfied and there existed no fear for the future development. Ten percent were just satisfied with the results. The complaints were expressed mainly because of the size of the penis and/or ugly scars.

In our material there was a good correlation between the appreciation of the surgeon (over 90% reasonable results) and that of the patients (90% satisfied with the results).

We think, that, if the operation is performed earlier, the normal appearance of the penis after operation can prevent many of the psychosexual disturbances. This can only be evaluated, however, after another decade has passed.

Conclusions and summary

The aim of this study is to analyse the results of an operative method for the correction of hypospadias, that fulfils all the demands, that we think should be met by such a corrective method. The operation should:

- a. give a complete reconstruction in one stage procedure.
- b. allow for a correction in the majority of the hypospadiac entity.
- c. give a cosmetic result, that hardly differs from the non-operated genitalia with preservation of its sensibility.
- d. allow a correction at an age at which the psychological sequelae are as minimal as possible.
- e. have a high reliability, with a complication rate of less than 20 %.

If we analyse the results of hypospadias correction with the inner preputial island flap technique with regard to the above mentioned requirements, we can summarize the following conclusions:

- a) In the primary cases in over 70 % a reasonable result was obtained in one session. Neither surgeon nor patient or his parents felt the need for any further corrective procedure. A very important role in this study played the learning process of the author, with a much higher complication rate in the beginning. Over the years 1984 and 1985 the complication rate dropped till just over 15 %. In the hypospadiac "cripple" group a reasonable result was obtained in over 64% in one session. This is a much better result, than can be obtained with two-stage corrective procedures. Apart from requiring a second operation, at least every fifth patient needed yet a third

correction, with a much less physiological appearance of the genitalia as result.

b) Many patients with severe forms of hypospadias, like the perineal and peno-scrotal forms, as seen in adreno-genital syndrome cases and in patients with severe enzyme disturbances, are included in this study. Also in these patients the inner prepuce island flap technique was found to be reliable and gave good results.

c) We have adopted as reference for the outcome of the corrective procedure the boy with a uncomplicated healed circumcision. The fact, that the prepuce must be sacrificed, may be a valid point for criticism (compared to the normal outlook), but almost all operative methods use the foreskin to compensate for the shortness of the ventral penile skin. The fact, that the urethral orifice is located in the normal area of the glans and its form and localisation allows a good undivided urinary stream, makes this method quite unique. Most other methods produce a subglandular position of the orifice, even in those cases in which early postoperative a good glandular position was seen. The sensibility tests show that there was no significant loss of sensibility.

d) Without increase of the complication rate the method described here, can be performed easily at the age of one year. Patients, who are operated before the onset of puberty, show an adequate growth of the operated area after puberty. Also follow-up studies show no stricture formation at the anastomoses and no recurrent chordee in five patients out of this study, who passed puberty. At this moment we can not proof that operation at an earlier age reduces the psychological sequelae of this congenital anomaly. The ultimate proof for this can be obtained only on the basis of longterm follow up studies in the forthcoming decades.

e)The low percentage of bad results (less than 6 %) in the group of the primary cases and the relatively high percentages (in just over 21 %) in the "cripple group" confirms the adage that corrective surgery can only be done properly at one occasion: the first time! Taking in consideration the bad situation before the operation in the hypospadiac "cripple group" and the high standards to which we refer to for the outcome of the result, the relatively good outcome in the "cripple group" shows the reliability of this operative procedure.

We think, that we have been able to show by this study that all the foramentioned demands are best met by the method, that we have described. This corrective procedure allows a chordectomy and the creation of a neo-urethroplasty out of a pedicled, vascularized flap of the inner-prepuce, in an one-stage procedure.

In chapter I the morphological characteristics of the hypospadiac entity are described.

The ectopy of the orifice, the lack of a frenulum and the asymmetrical raphe are described as intrinsic features of this malformation. The chordee, which is very often present in hypospadias is discussed in detail. Epidemiological data on incidence, heridity, maternal factors and associated anomalies are given. The morphogenesis of both the normal genitalia and the hypospadias form is described.

Current views on the aetiology of this malformation are discussed.

Chapter II reviews the operative methods used for the correction of hypospadias, which formed the base for currently used techniques. Those methods are described in more detail which form a part of still widely used "classical" two-stage method. Finally the most recently developed one-stage procedures are discussed.

Chapter III deals with the transverse inner preputial island flap technique. This method is described step by step together with the two mostly used supplementary surgical procedures for the total correction of severe chordee. The use of special suture materials and the characteristics of the used wounddressing are elucidated.

In chapter IV investigations on the vascularization pattern of the human penis are reported. The experimental studies show that the use of a vascularized island flaps from the prepuce is feasible because of its typical bloodsupply.

In chapter V the results of the hypospadias correction with the transverse inner preputial island flap technique are analyzed. The epidemiological data on the 115 patients operated with this technique are given. The per- and post-operative complications are described. The results of our study, in which in 82 out of 87 patients a good or fair result was achieved in the primary case group (94.3 %) and in 22 out of 28 patients in the "cripple" case group (78.5 %), are compared with the results in the literature for one-stage as well as for two-stage procedures.

Chapter VI deals with the possible psycho-social consequences of an operation of the external genitalia. These consequences seem to become more pronounced if the correction is performed on an older age. An evaluation of several longterm follow-up investigations is given and is related to the results of our investigations.

Based on these studies the optimal age for correction of hypospadias seems to be about the age of one year.

In Chapter VII the conclusions based on this study are formulated and a summary of this thesis is given.

De bedoeling van deze studie is een operatieve methode ter correctie van de hypospadie aan te geven, die naar ons inzicht het meest voldoet aan de eisen, die aan een dergelijke correctie gesteld kunnen worden.

Deze operatieve methode dient:

- a. in een zitting tot een volledige reconstructie te voeren.
- b. toepasbaar te zijn in een zo breed mogelijk scala der afwijking.
- c. met behoud van de sensibilliteit een cosmetisch verantwoord resultaat te geven, dat zo min mogelijk van het normale, niet geopereerde mannelijk genitaal afwijkt.
- d. op een leeftijd toepasbaar te zijn, waarop de psychische gevolgen voor de patienten zo minimaal mogelijk zijn.
- e. een hoge betrouwbaarheid te bezitten, met een percentage complicaties dat minder dan 20 % bedraagt.

Wanneer we de resultaten van de hypospadie correctie met behulp van een gesteelde eiland flap van het preputium binnenblad evalueren, kunnen de volgende conclusies getrokken worden;

- a) In de groep der zogenaamde primaire patienten werd in meer dan 70 % een bevredigend resultaat gehaald in één operatieve sessie. Daarbij werden geen verdere operatieve correcties, noch door de chirurg, noch door de patienten of diens ouders noodzakelijk c.q. wenselijk geacht. Daarbij dient bedacht te worden, dat het leerproces in deze studie een belangrijke rol speelde bij de auteur. Over het jaar 1984 en 1985 daalt het percentage noodzakelijke tweede ingrepen tot rond de 15 % . Bij de groep der zogenoemde "Hypospadias Cripples" werd in meer dan 64 % een bevredigend resultaat gehaald in één sessie. Dit is een, in vergelijk met de zogenoemde twee tijden technieken, die nog in meer dan 22 % een vervolg ingreep kennen, (buiten de altijd al noodzakelijke tweede ingreep), duidelijk beter resultaat.

b) In deze studie worden veel patiënten gepresenteerd met zeer ernstige vormen van hypospadie, zoals de perineale en peniscrotale vormen bij bijvoorbeeld het adrenogenitaal syndroom en bij ernstige enzymatische stoornissen. Ook bij deze patiënten gaf de eiland flap techniek reproduceerbaar goede resultaten.

c) Wij gaan er vanuit dat de enige standaard, waarmee het postoperatieve resultaat vergeleken mag en kan worden, een jongen is, waarbij de circumcisie probleemloos genezen is. Het feit dat de voorhuid geofferd wordt, mag een punt van kritiek zijn in vergelijking met de normale toestand, maar bij bijna alle operatieve correcties wordt de voorhuid tijdens de procedure, wegens het altijd aanwezige tekort aan huid aan de ventrale zijde, verbruikt. Het feit, dat het urethra ostium op de normale plaats op de glans is gesitueerd en door zijn vorm en lokalisatie een goede ongedeelde urinestraal geeft, maakt deze operatieve methode vrij uniek. Bijna alle andere methoden leveren een subglandulaire positie der meatus op, zelfs in die gevallen waarbij aanvankelijk postoperatief een glandulaire positie bestond.

Het sensibiliteits onderzoek liet zien dat met deze methode nauwelijks stoornissen optreden.

d) De beschreven operatie techniek kan even succesvol op vijf als op eenjarige leeftijd toegepast worden. Dit gaat niet gepaard met een toename van het aantal complicaties. De patiënten, die na een operatie vóór de puberteit, lichamelijk volwassen geworden zijn, vertonen een adequate groei der geopereerde gebieden. Tot nu toe is in de vijf patiënten die door de puberteit heen zijn geen strictuur van de anastomose plaatsen, noch een recidief verkromping der penis opgetreden. De grote voordelen betreffende de psychische consequenties zijn weliswaar reeds duidelijk zichtbaar, maar kunnen pas in de komende decaden, door longitudinale vervolg onderzoeksgegevens, wetenschappelijk onderbouwd worden.

e) Het geringe percentage slechte resultaten van minder dan 6 % in de voordien nooit geopereerde groep en van iets meer dan 21 % in de "Cripple" groep, onderschrijft nog eens duidelijk het adagium, dat correctieve chirurgie eigenlijk slechts één keer een goed resultaat kan geven, namelijk: de eerste keer! Mede gezien de voorheen al slechte situatie en de hoge eisen gesteld aan het benoemen van een resultaat als goed of tevredenstellend, geven deze cijfers juist bij de "Cripple" groep toch een hoge mate van betrouwbaarheid der methode weer.

Wij menen aan de hand van deze studie aangetoond te hebben dat aan de gestelde eisen het best door de door ons beschreven methode voldoet. Deze correctieve techniek maakt een chordectomie en de creatie van een neo-urethra uit een gesteelde eiland flap van het preputium binnenblad in één operatieve sessie mogelijk.

In Hoofdstuk I worden de morphologische kenmerken van de hypospadie beschreven. Intrinsiek aan deze malformatie zijn de ectopie van het orificium en de asymmetrie van de raphe. De zeer frequent voorkomende verkromming van de penis (chorda) wordt in detail beschreven. Begeleidende anomalieën en epidemiologische gegevens als incidentie, erfelijkheid en maternale factoren worden behandeld.

De ontstaanswijze van zowel de normale externe genitalia als ook van de hypospadie worden beschreven. De huidige inzichten in de oorzaak van deze malformatie worden besproken.

In Hoofdstuk II worden de operatieve correctie methoden geschilderd die basis vormen van de huidige technieken. Speciaal de nog veel gebruikte standaard twee-tijden technieken worden toegelicht. Tenslotte worden de nieuw ontwikkelde één-tijdse technieken beschreven.

Hoofdstuk III behandelt de transverse inner preputial island flap techniek, waarbij de operatie stap voor stap uit de doeken wordt gedaan.

Het speciale hechtmateriaal, het verband en de twee meest gebruikte methoden ter totale eradicatie van de chorda worden beschreven.

In Hoofdstuk IV wordt het onderzoek naar de vascularisatie van de penis bij mensen beschreven. Deze experimenten tonen aan dat het gebruik van volledig gesteelde huid flappen van het preputium, dankzij de bijzondere vascularisatie hiervan, mogelijk is.

In Hoofdstuk V worden de resultaten van de operatieve correctie van de hypospadie middels de transverse inner preputial island flap geanalyseerd. De epidemiologische gegevens van de bestudeerde groep van 115 patiënten worden behandeld. De per- en post operatieve complicaties worden beschreven.

De resultaten in deze studie worden vergeleken met de gegevens uit de literatuur over de één-tijdse zowel als de twee-tijdse technieken. Deze resultaten waren goed of acceptabel in 82 van de 87 patiënten (94,3 %) die tevoren nooit geopereerd waren (de zogenaamde primary cases) en in 22 van de 28 patiënten (78,5 %),

die meer dan eenmaal reeds gecorrigeerd werden (de zogenaamde "cripple case" group).

Hoofdstuk VI beschrijft de mogelijk psycho-sociale consequenties van een operatief ingrijpen aan het externe genitaal. Deze gevolgen lijken ernstiger te zijn naarmate de patienten ouder zijn. De gegevens van diverse na-onderzoeken worden vergeleken met de gegevens uit deze studie. Op grond van deze studie lijkt de optimale leeftijd om tot correctie van de uitwendige genitalia over te gaan rond de twaalf maanden te liggen.

In Hoofdstuk VII worden de conclusies, die aan de hand van deze studie getrokken worden, beschreven en een samenvatting van de Hoofdstukken sluit deze studie af.

Das Ziel der vorliegenden Untersuchung war es, eine operative Methode zur Korrektur der Hypospadie zu erarbeiten, die den Anforderungen welche an eine derartige Korrektur gestellt werden, am besten genügt.

- a. Die vollständige Rekonstruktion sollte in einer einzigen operativen Sitzung durchgeführt werden können.
- b. Bei einer breiten Skala von Anomalien anwendbar sein.
- c. Sie muss zu einem kosmetisch akzeptablen Ergebnis führen, welches möglichst wenig von dem normalen männlichen Genitale abweicht und die Sensibilität erhält.
- d. Die Operationstechnik muss in einem Alter möglich sein, in dem die psychischen Folgen der Operation gering bleiben.
- e. Sie muss zuverlässige Resultate mit einer Komplikationsrate unter 20 % liefern.

a) In der Gruppe sogenannter "primärer" Patienten konnten bei über 70 % der Fälle in einer einzigen Operationssitzung befriedigende Ergebnisse erzielt werden. Dabei wurden weder von den Chirurgen noch von den Patienten oder deren Eltern eine weitere chirurgische Korrektur für notwendig oder wünschenswert erachtet. Es sei allerdings auf den mit der Studie einhergehenden Lernprozess des Autors verwiesen. Im Verlauf der Jahre 1984 und 1985 schrumpft der Anteil notwendiger Zweitoperationen auf ca. 15 % . In der Gruppe sogenannter "Hypospadias Cripples" konnte bei über 64 % in einer Sitzung ein befriedigendes Resultat erzielt werden. Damit liegt die Erfolgsquote dieser Methode deutlich über der von zweizeitigen Korrekturen, die bei 22 % neben dem immer notwendigen zweiten Eingriff noch eine Folgeoperationen nach sich ziehen.

b) In dieser Studie wurden viele Patienten mit sehr gravierenden Varianten der Hypospadie versorgt, wie perineale und peniscrotale Formen z.B. beim androgenitalen Syndrom und anderen

schweren enzymatischen Störungen. Auch in diesem Patientenkollektiv wurden mittels der Präputium-Innenblatt-Lappen-Technik gute und reproduzierbare Ergebnisse erzielt.

c) Wir gehen davon aus, dass der einzige Massstab, an dem das postoperative Ergebnis gemessen werden kann, die unproblematisch verheilte Circumsion ist. Kritik mag an der Tatsache geübt werden, dass bei dem Eingriff die Vorhaut geopfert wird, aber beinahe alle operativen Rekonstruktionen verbrauchen das Präputium aus Mangel an Haut der Ventralseite. Diese Operationsmethode stellt insofern eine Besonderheit dar, weil das Ostium urethrae an seinem normalen Platz auf der Glans lokalisiert ist. Seine Form und Lage bedingen einen guten Urinstrahl. Fast alle andere Methoden führen zu einer subglandulären Position des Meatus, selbst bei den Techniken, bei denen postoperativ anfänglich eine glanduläre Lage bestand. Die Sensibilitätskontrolle konnte nachweisen, dass mit der von uns beschriebenen Technik kaum Störungen auftreten.

d) Die beschriebene Methode kann mit gleichem Erfolg sowohl im ersten als auch im fünften Lebensjahr, ohne Zunahme der Komplikationen angewandt werden. Patienten, die nach der, präpubertal durchgeführten Operation körperlich erwachsen wurden, zeigten ein adäquates Wachstum der operierten Anteile. Bislang sahen wir vor allem keine Strikturen in den Anastomose-gebieten, und auch ein Krümmungsrezidiv des Penis wurde nicht beobachtet bei den bis jetzt fünf Patienten, die bereits post-pubertal sind. Die grossen Vorteile in Hinblick auf die psychischen Folgen können, obwohl bereits jetzt deutlich nachweisbar, erst in den zukünftigen Jahrzehnten wissenschaftlich untermauert werden.

e) Die kleine Anzahl schlechter Ergebnisse unter 6 % bei Erstoperierten und etwas mehr als 21 % in der "Cripple"-Gruppe unterstreichen nochmals deutlich, dass Rekonstruktionschirurgie nur ein einziges Mal gute Resultate liefert, nämlich beim ersten Mal. Angesichts der ungünstigen Ausgangssituation und

der hohen Kriterien, ein Ergebnis als gut oder befriedigend zu beurteilen, spiegeln obige Prozentzahlen gerade in der "Cripple"-Gruppe doch eine grosse Verlässlichkeit der Methode wieder.

Wir glauben, dank der vorliegenden Untersuchung, mit der Chordektomie und Rekonstruktion einer Neourethra mittels eines gestielten, vaskularisierten Präputium-Innenblatt-Lappens in einer operativen Sitzung, die Methode gefunden zu haben, die die oben genannten Forderungen am besten erfüllt.

Kapittel I behandelt die typischen Hauptmerkmale der Hypospadie, wie die Ektopie des Ostiums urethrae und die Asymmetrie der Raphé. Die oft begleitende Krümmung des Gliedes (Chorda) wird im Detail beschrieben. Die Begleitmissbildungen und die epidemiologischen Aspekte wie die Inzidenz, die Heredität und die maternalen Faktoren werden abgehandelt.

Die Morphogenese des normalen externen Genitals sowie auch die des hypospaden Genitals werden beschrieben.

Der derzeitige Kenntnisstand der möglichen Kausalität dieser Malformation wird besprochen.

Kapittel II befasst sich mit den Operationsmethoden, die die Grundlage für die heutigen Techniken bilden. Vor allem die noch gängigen zweizeitigen Operationsverfahren werden wiedergegeben ebenso wie die neuentwickelten einzeitigen Techniken.

Kapittel III gibt die Korrektur mittels der Quere-Preputial-Innen-Lappen Technik detailliert wieder. Der Gebrauch besonderen Nahtmaterials und Verbandes wird erläutert. Die zwei meist angewandten komplementären Eingriffe zur endgültigen Chorda Korrektur werden beschrieben.

Kapittel IV gibt die Untersuchungsergebnisse des Vascularisationsmusters beim menschlichen Penis wieder. Diese zeigen, dass die spezielle Blutversorgung des Preputiums die Verwendung von gestielten Vorhautlappen ermöglicht.

Kapittel V gibt die Ergebnisse der operativen Korrektur mittels des Quere-Preputial-Innenblatt-Lappens unter Einbeziehung relevanter epidemiologischer Daten der 115 beschriebenen Patienten wieder.

Die prä- und postoperativen Komplikationen werden erläutert.

Die Ergebnisse werden mit relevante Literatur über ein- oder zweizeitig operierte Patienten verglichen.

Die Ergebnisse waren gut oder akzeptabel bei 82 von 87 Patienten (94,3 %), die nie vorher operiert worden waren (die sogenannte primary case Gruppe) und bei 22 von 28 Patienten (78,5 %), die mehrmals voroperiert waren (die sogenannte "cripple case" Gruppe).

Kapittel VI beschreibt die mögliche psycho-sozialen Folgen, die nach Operationen am äusseren Genitale altersabhängig auftreten können.

Einige Untersuchungen mit Langzeit-Follow-up werden bewertet und den Ergebnissen dieser Studie gegenüber gestellt.

Auf Grund dieser Studie erscheint das optimale Lebensalter für die operative Korrektur des äusseren Genitale rund zwölf Monate zu sein.

Kapittel VII beendet schliesslich mit den Schlussfolgerungen und der Zusammenfassung diese Promotionsarbeit.

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Jos de Vries was born on 15th December 1944 in Rotterdam. His secondary school certificate, Gymnasium B, was obtained in June 1964 at the St. Franciscus college Rotterdam.

From 1964 to 1972 he studied medicine at the Medical School of the Catholic University of Nijmegen. From 1966 to 1969 he was appointed research assistant in the department of physiology.

After receiving his medical degree in 1972 he served in the army in the eastern territoire as district psychiatrist.

From 1973 to 1976 he received his surgical training in the St. Johanner Teaching Hospital in Rheinhausen (Chief Dr. Med. G. Weitz) W. Germany.

The urological training followed at the St. Antonius Teaching Hospital in Eschweiler/Aachen (Chief Prof. Dr. Med. L. Steffens) W. Germany. In January 1979 he was appointed vice chairman of the department of urology in Eschweiler and in May 1979 he was registered as urologist in Germany.

In August 1979 he accepted the appointment as chef de clinique at the department of urology of the Catholic University Nijmegen.

His registration as urologist in the Netherlands followed in May 1980.

From 1980 to 1981 subsequent training in paediatric urology was received in Baltimore (R. Jeffs), Philadelphia (J. Duckett), Detroit (A. Perlmutter), Chicago (L. King) and Boston (H. Hendren) in the USA. Half a year supplementary training was obtained at the Hospital for Sick Children (Ph. Ransley) in London and at the Sophia Childrens Hospital (Prof. Dr. R. Scholtmeyer) Rotterdam.

In 1981 he was registered as paediatric urologist.

The 8th of Januari 1982 he opened the Paediatric Urology Centre (KUC) at the St. Radboud Hospital of the Catholic University of Nijmegen.

In October 1983 the workgroup on prenatal diagnosis and treatment was initiated.

He operated at the University Hospitals in Szezed (Hungary), Berlin, Bonn and Tübingen (W. Germany) and at major Teaching Hospitals in Berlin, Essen, Hörter and Osnabrück (W. Germany).

STELLINGEN

behorend bij het proefschrift

HYPOSPADIAS REPAIR WITH THE TRANSVERSE INNER PREPUTIAL ISLAND FLAP TECHNIQUE

12 september 1986

JOSEPH D.M. de VRIES

1. De indicatie tot operatieve correctie van een hypospadie wordt mede door de psychische consequenties ervan gesteld.
2. Volgens de huidige inzichten in de ontwikkeling van het kind is de periode rond het eerste levensjaar het meest geschikt voor de correctie van het externe genitaal.
3. Bij de correctieve chirurgie van het externe genitaal dient niet alleen de huid fraai gesloten, maar ook een urethra gevormd te worden, die toekomstige groei toelaat.
4. Caudaal anaesthesie, door middel van langwerkende lokaal-anaesthetica met optimale sensibele blokkade tijdens en langdurige pijnstilling na scrotale en peniele chirurgie, zou meer moeten worden toegepast.
5. Het gegeven dat veel van de hypospadie-patiënten reeds meerdere malen zijn geopereerd, zegt niets over de moeilijkheidsgraad van de correctie der aandoening, maar meer over de onervarenheid van de behandelaren.

6. Bij het algemeen lichamelijk onderzoek van een zuigeling mag het meten van de bloeddruk niet worden nagelaten.
7. Het onderzoek naar de vorm van de erythrocyten in de verse urine, levert ook bij het kind een belangrijke bijdrage tot de localisatie van de haematurie.

Rizzoni G., Braggion F., Zacchello G.
'Evaluation of glomerular and non-glomerular Hematuria by Phase-contrast Microscopy'
J. Pediatr.; 103, 370-374, 1983.

8. De correctie der maldescensus testis dient vóór het tweede levensjaar te geschieden.

Hadziselimovic F., Herzog B., Girard J., Stalder G.
'Cryptorchidism: Histology, Fertility and Treatment'.
In: Progress in reproductive Biology and Medicine.
Ed. Kenouim A., Pryor J.B.
S. Karger Basel, p. 1, 1984.

9. Een kind met secundaire enuresis, met op het röntgenbeeld een niet adequate sluiting van één of meer wervelbogen, dient urodynamisch onderzocht te worden ten einde een eventueel 'spinal tethered cord' zo vroeg mogelijk aan te tonen.
10. Kinder-Urologie is geen 'volwassen' Urologie bij kinderen, maar een vakgebied, dat zich bezighoudt met en zonedig correctief optreedt in het zich in ontwikkeling bevindende urogenitale systeem.

11. Incontinentia urinae bij de vrouw is geen afwijking van de vrouwelijke genitalia, maar een stoornis in de urodynamische karakteristieken van de blaas en de urethra.
12. Diagnostische beeldvormende middelen, zoals de echografie, niet gebruik makend van ioniserende straling en radio-actieve stoffen, dienen ook door een orgaanspecialist te kunnen worden ingezet en in therapeutisch handelen te kunnen worden omgezet.
13. De invoering van een toetsing van de aktuele vakbeheersing van de 'zittende' artsen is van belang voor het peil van het geneeskundig handelen.
15. Milieubeheer zal pas haar juiste dimensie verkrijgen, indien de mensen de aarde niet langer als 't erfgoed hunner vaders, maar als een lening voor de toekomst van hun kinderen beschouwen.
16. Een aangeboren afwijking komt nooit alleen, er zit altijd een kind aan vast.

